POSTER PRESENTATIONS

PP001 - THE EFFECT OF BREAST MILK ON THE HEALING OF DIAPER DERMATITIS

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Background: Diaper dermatitis is the most common dermatological disease of infancy, occurring and is caused by the combined effect of irritants such as diaper, urine and feces.
Objective: In this study we want to evaluate the effect of breast milk on the healing of diaper dermatitis.
Methods: The present study is a clinical trial which is done on Health Centers of Tehran in Iran(2007).Samples studied include 30 infants between 0 – 12 months of age that suffering from diaper dermatitis. Infants who did not respond within 5 days were given conventional therapy and the failure was recorded.Infants were divided into two matched groups: case and control, they were asked to clean the infant following urination /defecation with warm water and dry him/her .The case group was asked to apply the breast milk on the lesions for 5 days.Data was collected by a questionnaire containing two parts: the demographic characteristics of infants and the status of care and condition of the lesion according to Al-Waili study(1). Statistical tests (chi – square test, fisher test, Mann- Whitney test) were used to analyze the data.
Results: Findings showed significant differences between the case and control groups in the number and lesion score of the rashes at the first and third day (p=0.013),(p=0.005) .These differences was more significant at the fifth day (p=0.004), (p=0.001).
Conclusion: This study reveals that breast milk significantly decreases the number and lesion score of dermatitis and has a healing effect on infants' diaper dermatitis.
Key words: Breast Milk, Healing, Diaper Dermatitis.
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PP002 - VERNIX CASEOSA : AN HYDROPHOBIC LAYER

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Introduction: Well before birth, the fetal skin is against the external environment (amniotic liquid) by a specific layer called Vernix Caseosa (VC). This protection begins towards the 24th gestation week and is made of a layer containing up of 10% protein, 10% lipid and 80% water.(stratum corneum contains only 12- 32% of water). This unique complex proteolipid human material is composed of sebum, epidermal lipids, and desquamated epithelial cells.
This layer plays fundamental roles such as: barrier to water loss, temperature regulation, innate immunity. Its great water capacity can hydrate the skin of newborn and helps it to adapt to dry environment. Moreover it is involved in the acid mantle formation. The water contact angle qw is an indicator of the surface hydrophobic/hydrophilic tendency.
Objectives: The objective of this work was to quantify the hydrophilic/hydrophobic tendency of VC in order to compare it with in vivo human skin wettability.
Materials and methods: The water contact angle qw was measured on the fleshly VC by a tool specially conceived for in vivo and/or in vitro wettability studies.
Results: The water contact angle qw was about 88°. This value classed VC within hydrophobic tendency surfaces.
Discussion: In this work, our results were closed to those in the literature. The values classed VC as a surface of hydrophobic tendency compared to that measured on the volar forearm (poor in sebum). In the forehead (rich in sebum) the qw = 55°. Conclusion and Perspectives: However, in order to best understand the exact nature, role and for future exploitation of this material, both faces not yet studied till today, should be tested. The bibliographic data show that VC material may be used in vivo for dry skin hydration and could help in anti microbial and innate immunity defense.
Key words : Vernix Caseosa, wettability, water contact angle, surface energy, Hydrophobia.

PP003 - DIFFERENCIES BETWEEN ERYTHEMA TOXICUM AND ACNE NEONATORUM IN BOSNIA AND HERZEGOVINA

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Background: Erythema toxicum neonatorum and transient neonatal acne are among the many benign skin conditions that can occur in newborns.
Objectives: Estimation ethnics and sex differencies between newborns in Bosnia and Herzegovina.
Methods: The rate of two skin disorders and their relationship to ethnic member population and sex were calculated and analyzed using the computerized program Sigmastat 3.0.
Results: It has been estimated that up to 13% of newborns in Bosnia may be affected with neonatal acne and up 33% to erythema toxicum. Neonatal acne typically peaks at about age 3 months and rarely requires treatment. Of the 92 patients studied, 62 were male neonate (67.4%) and 30 were female (32.6%). Neonatal acne occurs in about 22.5 % and erythema toxicum neonatorum in about 55% of newborns in Gypsies ethnics neonatal population.
Conclusions: Benign dermatoses in newborns must be distinguished from more serious disorders with cutaneous manifestations. No identification differencies beetween sex, ethnics and religions neonatal population in skin neonatal acne and erythema toxicum neonatorum except orginal Gypsies population with higher rates comparing other ethnics groups in Bosnia and Herzegovina, but similiar rates with other European groups and rates.
References
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PP004 - CONGENITAL LESIONS WITH THE HAIR COLLAR SIGN

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Introduction: The hair collar sign is a ring of long and darks hair surrounding a congenital scalp lesion, usually composed of heterotopical neural tissue. Image studies prior to biopsy or excision are perceptive.
Cases report: We report 4 patients with congenital lesions with the hair collar sign. Image studies showed no intracranial communication in patients 1 to 3. Patient 1. A 1-day female showed a well-circumscribed, soft, exophytic yellow mass over interparietal area, surrounded by a ring of dark large hair. Histological analysis resulted of nevus sebaceous. Patient 2. A 7-day female showed 3 rounded, sharply marginated, depressed and hairless plaques in a linear disposition over her right parietal area, with hair collar sign. They were diagnosed as membranous aplasia cutis Patient 3. A 6-month-old boy presented a persistent hairless plaque on his left parietal area since birth, with a ring of hair longer and darker than the rest, consistent with membranous aplasia cutis. Patient 4. A 10-day male showed a nodular, skin-coloured nodule with cartilaginous texture on the right side of his neck, surrounded by a ring of dark hair. A cervical echography showed no cyst nor fistula. Clinical diagnosis was congenital cartilaginous rest of the neck.
Discussion: Nevus sebaceous is a congenital lesion characterized by an excess of glandular skin structures. Presentation as an exophytic mass with a hair collar sign is extremely unfrequent. Membranous aplasia cutis is a group of disorders in which areas of the skin are absent or scarred at birth. It can be a failed expression of neural tube closure defect, as well as the hair collar sign. Congenital cartilaginous rest of the neck is considered a cervical variant of accessory tragi. To our knowledge, hair collar sign has not been previously reported in this entity, and could be related with developmental anomalies of ectodermal structures.

PP005 - A DECRIPTIVE STUDY OF DEMOGRAPHIC CHARACTERISTICS IN INFANTS WITH DIAPER DERMATITIS

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Background: Diaper dermatitis commonly affects infants, with peak incidence occurring when the individual is aged 9-12months. Studies determined that at any given time, diaper dermatitis is prevalent in 7-35% of the infant population. Diaper dermatitis is caused by over hydration of the skin, maceration,prolonged contact with urine and feces.(1)
Objective: In the present study we descript demographic characteristics in infants with Diaper Dermatitis.
Methods: Fifty infants between 0-12 month ages who suffering from diaper dermatitis,were studied. At baseline,a through medical history was obtained and a complete medical examination was conducted. Nobody has Diarrhea &antibiotic therapy. Data was collected by a questionnaire that is filled through interview with their mothers.
Results: Most of infants had the first birth seniority. All the mothers were house wives and 53.3% of the fathers had privet jobs.30% mothers and 33.3% fathers had (a qualification of) Diploma or higher. The mean ages of mothers were 25.44 years & mean ages of fathers were 30.5.All infants were used high power absorbent material for diapering.The most common place of dermatitis rash was Anal, although Genitalia & perineum were involved too. No sexual difference was observed. Diaper rashes were most common between 8-10 months old. 73.3% infants have one site of Diaper rash. Also 60%infantes have moderate rash.
Conclusion: Diaper irritant contact dermatitis is the most prevalent diaper dermatitis and,probably,the most common cause of coetaneous disease in infants.Diaper rashes can occur intermittently,anytime while your child wears diapers.
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PP006 - COMPARISON OF USE OF BREAST MILK (HBM)ON DIAPER RASH BETWEEN IRANIAN AND AFGHAN MOTHERS IN TEHRAN(2009)

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Background: A diaperrash isan areaofinflamedskin found in the diaperarea ininfants.Itis usually caused byskin irritation from prolonged contact with urineandfeces.Purpose:In this study ouraim wasto determine the effectof human breastmilk on diaperrash in infant's of Iranian and Afghan mothers.
Methods: 62 infants between 0-12months ofage that suffering from diaperrash and coming to our clinics were selected.The first and second part of questionnaire are demographic characteristics of the mothers and their infants.The third part is about the status of using breastmilk during7days.Descriptive analytical statistical tests onafive–pointscale. Both samples were asked to clean the infants after urination or defecation with warmwater,then drythe infant and rub breastmilk on diaperrash at least5times aday and for7days.
Results: Findings showed that the most common places of diaper rash in day first is genitalia, perineum and anal and by using breast milk the incidence of anal diaper rash has decreased. Both groups had the same diaperrash by place. At the beginning of study, 76-82% of infants had score 2 and by day 3, 87% of them got score 1. Results following our goal, showed in the day 3 were no differences between two groups of infants by number or rashes. In day 7, there were norashin 91% of infants.
Conclusion: The study treatment was effective in reducing

the symptoms of diaper dermatitis. The mother breast milk were aneffective treatment for diaperrash in twogroups. Since nature designed human milk for human babies, it is the most easily digested drugs that baby can received for his napkin rash.

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PP007 - APLASIA CUTIS CONGENITA IN ASSOCIATION WITH FETUS PAPYRACEUS

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Introduction. Aplasia cutis congenita (ACC) is a rare disorder characterized by localized absence of skin at birth. In cases of truncal or extremity ACC the condition is often related to the death of a co-twin in utero. We describe two cases of ACC in association with fetus papyraceus.

Case reports. Case 1: A girl was delivered by caesarean section at 41 weeks gestation due to placental abruption. At birth widespread ulcerative skin defects were seen. Furthermore a mummified dead twin - fetus papyraceus – about 10 cm long was found. With conservative skin care the defects healed in 6 weeks. At age 7 months large, symmetrical, cicatricial and sclerotic skin areas were found in an H-shaped pattern on the back and legs while changes were more circular on the abdomen (Pic). Case 2: A boy was delivered by a planned caesarean section at 38 weeks gestation. He was the result of a triplet pregnancy conceived through microinsemination, consisting of one unaffected dizygotic triplet and two monochorionic twins of whom one spontaneously died in week 20 and was found at birth as a fetus papyraceus. At birth macular, atrophic, slightly red skin changes symmetrically on the legs were found, changing over time to hypopigmented, sharply defined macules (Pic).

Discussion: Our two cases represent completely different clinical presentations of ACC in association with a fetus papyraceus. The mechanism for this rare type of ACC may involve acute ischemia of "watershed" areas of the skin as a result of sudden relaxation of the vascular bed of the dying, monochorionic, twin, leading to hypovolemia in the surviving twin. The differing severity in the two described cases may reflect differences in the gestational time of death of the co-twin.

PP008 - PSEUDOAINHUM CAUSED BY HAIR TOURNIQUET SYNDROME: REPORT OF TWO CASES

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Pseudoainhum is a term used to describe the presence of constricting bands of the extremities due to a variety of underlying causes. The hair-thread tourniquet syndrome, or toe tourniquet syndrome, occurs when hair, and occasionally thread or fiber, wraps tightly around a young child's appendage and obstructs the circulation. Progression of the lesions can cause irreversible damage and autoamputation of the affected digit. This syndrome has been described to involve the fingers, the toes, and even the genitals. The offending fiber can cut through the skin making the fiber difficult to see. The standard treatment is surgical, with prompt removal of the constricting fiber. Although most cases are felt to be accidental, child abuse must be considered in selected cases.

The authors report two cases of hair tourniquet syndrome, in two healthy infants of 2 and 3-months-old, affecting the fourth and the fifth toe respectively. In both cases the toes were edematous and very painful, and it was observed a circumferential groove caused by the constricting hair, but there were no signals of ischaemia or tissue necrosis. Removal of the hair was achieved in both cases. The authors stress the importance of recognizing this rare condition and of prompt, complete, surgical release.

PP009 - COMPARING THE EFFICACY OF TOPICAL SUCRALFATE VERSUS TOPICAL ZINC OXIDE IN DIAPER DERMATITIS, A RANDOMIZED, DOUBLE BLIND STUDY

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Background: Diaper rash is not a life threatening condition, but may affect quality of life in both young children and their parents.Sucralfate is a basic aluminum salt of a sulfated disaccharide, which makes a physical barrier and accelerates ulcers healing. The aim of this study was to compare the efficacy of topical sucralfate versus topical zinc oxide in diaper dermatitis.

Method: In this randomized, double blind study, sucralfate and zinc oxide were formulated as 20% ointments with same excipients.

After passing all needed tests such as stability, pH, hemogenicity and microbial tests, two formulations were coded and dispensed to the pediatricians.

46 inpatient infants and young children age 1-24 months were enrolled in this study. Informed consents were obtained from the parents as ordered by ethics committee.

All patients were treated topically with either sucralfate (N=25) or zinc oxide (N=21) for 7 days.

Diaper severity scores were obtained before treatment and at days 3, 5, 7.

Results: After termination of the study and decoding, it was shown that sucralfate 20% ointment was significantly superior in healing diaper dermatitis at days 5,7($P<0.05$ and $P<0.01$ respectively) in comparison with zinc oxide 20% ointment.

Also complete healing time in sucralfate group (3.24±2.02 days) was significantly shorter than zinc oxide group (5.42±2.39days).

Tolerability was excellent in two groups and no adverse effects were reported in either group.

Conclusion: Since sucralfate in topical formulations acts as a physical barrier with proved safety profile and has no noticeable absorption, it may become a potential treatment for diaper dermatitis.

PP010 - SKIN CARE OF PHOTOTHERAPY IN NEWBORN BY USING BREAST MILK (HBM) IN TEHRAN (2009)

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Background:The skin of newborns during phototherapy have special needs for protection from dermatitis, brons baby syndrome. Turning of infants during phototherapy and hyperbilirobinemia is practiced in many nurseries. This study examin the effect of breast milk during photography among 3-7 days of age's newborns study design.
Methods: we first choused 32 newborns in twe groups. The one whose mothers rubbed breast milk on her baby's skin,and the other did not.
Result: Thirty term infants were in this study.no differences were found between the groups in baseline data such as birth weight age. Infants in wich used breast milk, showed no signs of dermatitis and brons baby, and in the other 15 infants there were 3 brons baby, 5 dermatitis, and 6 diaper rash. Turning the infants were the same and the level of bilirobin in bith groups were 11.2- 15.7. The mothers used breast milk 5-7 times a day.
Conclusion. We conclude that breast milk should be used to prevent infants from dermatitis and to treat them.Since nature designed human milk for human babies, mother's milk is the most easily digested drugs that baby can received for.It also helps prepare a baby's immune system and skin protect against sensitivity specialy during phototherapy.
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PP011 - SYSTEMATIZED POROKERATOTIC ECCRINE OSTIAL AND DERMAL DUCT NEVUS IN A NEWBORN SHOWING PARTIAL REGRESSION THREE MONTHS LATER

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An one-day-old healthy girl presented with erythematous keratotic papules following Blaschko's lines in a symmetric fashion on the whole body. The papules coalesced into verrucous plaques on the sacrum and the dorsa of hands and feet. Three months later the streaks had disappeared and the plaques had flattened. A biopsy from the rest of the sacral plaque revealed epidermal invaginations containing cornoid lamellae exclusively involving the acrosyringia. The diagnosis was consistent with a porokeratotic eccrine ostial and dermal duct nevus (PEODDN).
PEODDN is a rare disorder of keratinisation reflecting cutaneous mosaicism. It mostly occurs on the palms, soles and distal extremities. Eight cases of systematized PEODDN have been reported and only one patient showed incomplete involution within 22 years later. Because of the unusually fast regression in our patient, a mutation of the NEMO-Gene was excluded. Differential diagnoses, histogenesis and possible causes for the fast regression are going to be discussed.

PP012 - INCOMPLETE DEVELOPMENT OF THE HALLUX NAIL IN THE NEWBORN

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Between March and October, 2008 we observed in the Unit of Neonatology the nails of the hands and feet of 541 (252 females) consecutively born neonates at an average age of 3.2 days. Moreover, we visited again 36 of these newborns with nail alterations 2-6 months later.
The most frequent alteration was the incomplete development of the hallux nail, appearing as a triangular or trapezoidal nail. This alteration had been previously described in the relevant literature as congenital hypertrophy of the distal and lateral nail folds (1). It was observed in 347/541 newborns,but disappeared spontaneously within 1-3 months. It was never associated with inflammation or onychocriptosis. The seeming hypertrophy of the nail folds was not real, but secondary to the lacking pressure of the nail lamina on themselves. The incomplete development of the hallux nail is probably due to an asynchronic growth of the nail lamina with the nail cutaneous folds.
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PP013 - BLUEBERRY MUFFIN BABY DISCLOSING CONGENITAL CYTOMEGALOVIRUS INFECTION: CASE REPORT

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Case report: A male caucasian neonate was born at 37 weeks gestational age following a pregnancy complicated by polyhidramnios. He was small for age and skin examination revealed a generalized eruption of blue-red dome-shaped papules and nodules favoring the head and extremities. No other changes were notable on physical exam except for hepatosplenomegaly. The histology of a skin nodule showed dermal erythropoiesis with the presence of mature erythroblasts. The laboratory tests revealed thrombocytopenia, direct hyperbilirubinemia, a negative Coombs test and the bone marrow aspirative biopsy was normal. The PCR assay of blood specimen was positive for cytomegalovirus(CMV) with 2,2x103 copies/ml and negative for the rest of the TORCH infections. Both mother and child serum were positive for IgG anti-CMV but negative for IgM. The placenta presented histologic and immunohistochemical changes of CMV infection. The neonate received Ganciclovir treatment. At 2 months follow-up he was free of skin lesions.
Discussion: Blueberry muffin baby is a rare neonatal skin disorder characterized by red-blue macules, papules and nodules representing extramedullary hematopoiesis and is

associated to congenital infections and hemolysis. The descriptive term also includes several neoplastic and vascular processes. Skin biopsy must be performed to rule out these disorders. CMV congenital infection is the leading infectious cause of mental retardation, sensorineural deafness and visual impairment and the diagnosis during pregnancy is still complex. Antiviral treatment prevents hearing loss and development delays.

PP015 - PHACOMATOSIS SPILOROSEA WITH SYSTEMIC INVOLVEMENT

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Case report: A 7-year-old ecuadorian child was referred to the clinic for evaluation of an extensive pigmented and reddish patches of the skin, present since birth. There was no family history of similar lesions.
On phisical examination there were large areas of light-brown patched with numerous small (1-4 mm) darkly pigmented macules, located on the face, left arm and anterior and posterior aspects of the upper trunk, with a sharp demarcation respect the surrounding normal skin. A similar but smaller lesion was localized on the lumbar region.
A pale pink giant telangiectatic nevus (nevus roseus) was present on the left leg, and also on the left upper chest and the left arm. On chest and arm locations pigmented and red patches overlapped.
He presented dental malformations in position and size, with a reduced number of teeth. The nails were normal. There was no diference between the circumference of the extremities but the patient's left leg measured 1cm longer than the right leg, which resulted in scoliosis.
Three punch biopsies were taken. The biopsy from the preesternal region showed findings consistent with nevus spilus. The biopsy taken from the left leg revealed a dilatation of the superficial vessels. On the left arm we could see both lesions appearing together.
Neurologic and ophthalmologic examinations were unremarkable.
Summary: Phacomatosis spilorosea is characterized by the coexistence of a nevus spilus of the macular type and a telangiectatic nevus of a "nevus roseus" type.
It is an infrequent entity, with only 11 cases reported in the literature, the majority in japanese population. All of the cases are sporadic, except for two brothers published by Suzuki. The rest of the cases are isolated reports.
The disorder may occur with or without other anomalies associated. Abnormalities of the central nervous system, the eye, and the skeleton are the most frequent systemic complications. The most common associations of phacomatosis spilorosea are Sturge-Weber syndrome, Klippel-Trenaunay syndrome, and asymmetrical length of legs resulting in scoliosis.

PP016 - PSEUDOXANTHOMA ELASTICUM IN SIBLINGS AGED 7 AND 10 WITH SIGNS OF CALCIFICATION OF RENAL ARTERIES - CASE REPORT

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Background: Pseudoxanthoma elasticum (PXE) is a genetic disorder of the elastic tissue related to mutations in the ABCC6 gene. It involves the skin, eyes and cardiovascular system. The diagnosis is most often made in the second or third decade of life. Skin lesions may develop earlier in childhood, but may be easily overlooked. In childhood period, systemic signs of PXE in siblings are rarely described.
Methods: A case of the disease in 10 year-old girl an her 7 year-old brother is reported. Both presented with similar skin lesions on their necks. The first skin lesions were noted by their parents at the age of 6.
Results: At the examination at our clinic both children presented with soft, yellowish, xanthoma-like polygonal plaques resembling plucked chicken skin on their necks. Histopathological findings showed an increase in elastic tissue, which was clumped and fragmented in the middle dermis. In order to exclude systemic involvement, several diagnostic procedures were performed. Ophtalmological and cardilogical findings were unremarkable. In both of them abdominal ultrasound showed early signs of calcification of the intimal part of renal arteries. Low-fat diet and low intake of calcium were introduced in the therapy. Both are regulary followed up by a nephrologist, cardiologist and ophtalmologist.
Conclusions: Pseudoxatnthoma elasticum with early systemic signs is rarely discribed in siblings in childhood period. Behind discrete skin lesions which can be easily overlooked, there can be systemic involvement. Early diagnosis and careful checking for systemic signs is important and may improve the prognosis of the disease.

PP017 - KID SYNDROME:RECALCITRANT SCALP INFECTION WITH PROGRESSIVE SCARRING ALOPECIA

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Kid syndome is a rare congenital ectodermal disorder characterized by progressive vascularizing keratitis, decrease in visual acuity and blindness, non progressive deafness and icthyosis with features of erythrokeratoderma. It seems to be autosomal dominant disorder with mutation in the connexin 26 gene(GJB2).
Epithelial dysfunction of hyperkeratotic and underlying immune defect make these patients vulnerable to recurrent mycotic and bacterial infection and development of squamous cell carcinoma .
Here we report a 14-year-old boy, born of uneventful pregnancy and delivery from a non consanguineous parent who presented with classical features of Kid syndrome and recurrent episodes of scalp infection in last two years leading to sever progressive scarring alopecia.
Ophthalmologic examination disclosed significant photophobia, conjunctival injection with vascularizing keratitis and audiometric examination demonstrated sensorineural hearing loss in both ears. Skin examination revealed icthyosis like hyperkeratosis, reticulated palmoplantar keratoderma and sparse eyebrows. Scalp hair was sparse in occipital area with multiple crusted surface erosive plaques and pustules with foul odor discharge and sever atrophic scarring alopecia in remaining part of scalp accompanied with bilateral cervical lymphadenomegaly. Culture from scalp wound discharge

showed Candida Sp. and Proteus Sp. growth and primary investigation of immune system didn't reveal a common underlying systemic immune defect.
We think that along with major ophthalmologic and otolaryngologic problem susceptibility to recurrent skin infection needs urgent attention. Skin care and early detection of infection and initiation of antibiotics as well as for Candida should be considered in mind to prevent this associated morbidity. Further extensive investigation recommended to know underlying problem in their immune system.

PP018 - LETHAL JUNCTIONAL EPIDERMOLYSIS BULLOSA WITH PYLORIC ATRESIA DUE TO COMPOUND HETEROZYGOSITY FOR TWO NOVEL MUTATIONS IN THE INTEGRIN BETA4 GENE

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Junctional epidermolysis bullosa with pyloric atresia (JEB-PA) is a rare autosomal recessive disease with blister formation within the lamina lucida due to mutations in the integrin β4 (ITGB4) or integrin α6 (ITGA6) gene.
A girl, first child of healthy non-consanguineous parents, was born at 26 weeks of gestation by caesarean section, following polyhydramnion and abruption of placenta. She presented with extensive areas of denuded skin on head, neck and extremities. Auricles were hypoplastic. Abdominal ultrasound and X-ray were suggestive of pyloric atresia which was confirmed by laparotomy and surgical revision on the 4th day of life. Further course was complicated by progressive skin detachment, sepsis, and renal insufficiency with fatal outcome at 18 days of age.
Immunofluorescence mapping of cryopreserved skin showed junctional cleft formation with negative staining for integrins α6 and β4. Mutational analysis revealed compound heterozygosity for 2 novel mutations in ITGB4, both resulting in premature termination codons: c.600dupC/p.F201fsX14 and c.2533C>T/p.Q845X. Two subsequent pregnancies were terminated following prenatal diagnosis disclosing the same ITGB4 mutations.
In conclusion, we describe a case of lethal JEB-PA with negative immunoreactivity to integrins α6 and β4 being predictive of a poor outcome. Identification of compound heterozygosity for 2 novel ITGB4 mutations in the affected newborn enabled the prenatal diagnosis in 2 further pregnancies and exclusion of the disease in another pregnancy.

PP019 - HEREDITARY MUCOEPITHELIAL DYSPLASIA IN TWO GIRLS

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Introduction. Hereditary mucoepithelial dysplasia (HMD), is a rare genodermatosis characterized by nonscarring alopecia, fiery red gums, psoriasiform perineal lesions and ocular abnormalities (1,2). Histological studies show a psoriasiform epidermal hyperplasia with dyskeratotic keratinocytes. HMD displays an impaired epithelial cohesion in which no genetic origin has been elicit yet. Ultrastructural studies demonstrate epithelial dyshesion and a reduced number of desmosomes around the dyskeratotic cells, prompting thoughts that HMD could be a disorder of desmosomes and gap junctions. Differential diagnoses include keratitis-ichthyosis-deafness (KID) syndrome, ichthyosis follicularis-alopecia-photophobia (IFAP) syndrome and keratosis follicularis spinulosa decalvans (KFSD).
Case reports. We report on two unrelated girls of 10 and 16 years of age respectively who presented with typical clinical and histological features of HMD
Discussion. A diagnosis of HMD should be considered when facing the triad of non-scarring alopecia, well-demarcated redness of the oral mucosa and psoriasiform perineal rash, after exclusion of the clinically related follicular keratosis syndromes.
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PP020 - A SURVIVING CASE OF HARLEQUIN ICHTHYOSIS

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A baby, born at 36 weeks of gestation to consanguineous parents – their third child - with no past history of genetic skin disorders was found to be encased in a rigid, yellow-brown, hyperkeratotic, thick adherent "cast". He had marked ectropion, conjunctival oedema, eclabium and distorted facial features. He had hypoplastic ears, oedematous clenched hands and restricted movements. Deep pink fissures developed from day one but he was not distressed and had a strong and effective sucking reflex. He was transferred to a neonatal unit (nursed in a cot) and started intensive treatment with emulsifying ointment every three hours (500 grams every 2 days) with tubifast garments; regular lacrilube eye ointment and dermol 600 as a bath emollient. Fluids were given through an umbilical catheter and infantrini milk via a special teat.
Acitretin at a dose of 1.3 mg (0.5 mg per kg) was commenced on day 3 and this was adjusted according to weight. His skin gradually improved with shedding of the thick adherent scale leaving an erythrodermic background with scaling resembling severe non-bullous ichthyosiform erythroderma. He was discharged aged 4 weeks and has continued to make good progress since. A trial period off acitretin at 6 months led to gradual worsening of the hyperkeratosis and ectropion. He is now 20 months old and is on continuous therapy with good response. This case demonstrates the importance of early intensive topical therapy and nursing support combined with acitretin to improve the chances of survival.

PP021 - GORLIN-GOLTZ SYNDROME DIAGNOSED USING DERMOSCOPY IN A 2-YEAR-OLD GIRL

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Introduction. The Gorlin syndrome is an autosomal dominant condition caused by a germline mutation of the tumor suppressor gene PTCH1 on chromosome 9q22.3-q31. It is characterized by early onset of multiple tumors, including basal cell carcinomas (BCC) and medulloblastomas, odontogenic keratocysts, palmar and plantar pits, intracranial ectopic calcification and skeletal anomalies. Most cases present in the first and the second decades of life.
Clinical case. A 2 year-old-girl with hydrocephaly, frontal bossing and polydactily presented since birth three pigmented lesions on the dorsum of the left foot and fingers. On dermoscopy these lesions showed maple leaf-like areas, and multiple brown-grayish dots and globules. The histopathology of the lesion of the foot confirmed the diagnosis of a pigmented BCC.
Discussion. BCC in children younger than 15 years is rare, and is usually associated with a predisposing genetic condition such as Gorlin syndrome or xeroderma pigmentosum. More than 100 clinical abnormalities have been reported in Gorlin syndrome being BCCs the most characteristic finding. In our case, dermoscopy was a very useful tool for the correct diagnosis of the lesions as BCCs and aid in the diagnosis of the genetic syndrome.
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PP022 - SQUAMOUS CELL CARCINOMA IN PATIENTS WITH EPIDERMOLYSIS BULLOSA: REPORT OF THREE CASES

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Cutaneous squamous cell carcinoma (SCC) is the most significant and serious complication in patients with inherited epidermolysis bullosa (EB).
We present three patients with recessive dystrophic severe generalized EB (previously called Hallopeau Siemens) with cutaneous squamous cell carcinoma. In all three patients the tumours developed on distal parts of the limbs, that were exposed to chronic nonhealing erosions and ulcerations, which were finally severely mutilated. One patient had two primarily sites of SCC, the dorsum of the foot and the sacral region. No regional or distant metastases were detected.
All patients had surgical treatment; in two patients partial limb amputations and in one patient excision of the tumour with wide surgical margins were performed.
Cutaneous squamous cell carcinoma is predominantly seen in patients with dystrophic epidermolysis bullosa (DEB). The prevalence of SCC in recessive DEB Hallopeau Siemens is 23% with the cumulative risk of 90% by 55 years of age. These tumours have aggressive and fast grow and metastasise early representing the leading cause of death in these patients. Multiple appearance of SCC in EB patients is often observed. Early age of development is a rule and it has been described as early as 14 years of age. Several biopsies may be required to confirm or exclude the presence of tumour within a suspicious area.
Surgical treatment is the treatment of choice for SCC with amputations that are often indicated. Regular follow-up of non-healing wounds with early recognition of SCC is mandatory for effective treatment in patients with epidermolysis bullosa.

PP023 - PRIMARY HYPERTROPHIC OSTEOARTHOPATHY: AN UNUSUAL CAUSE OF EARLY-ONSET PALMOPLANTAR HYPERHIDROSIS

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Background: Primary hypertrophic osteoarthropathy (PHO) is a rare genetic syndrome characterized by periostosis, digital clubbing and pachyderma.
Methods and Results: A 12-year-old boy presented with congenital palmoplantar hyperhidrosis and arthralgias. Examination revealed thickening of the palmoplantar skin with clinically overt hyperhidrosis, hippocratic nails, and a cylindrical shape of arms and legs. Palmoplantar sweat production was threefold elevated in gravimetry, correlating to high sweat gland density on histology. Laboratory testing showed highly elevated serum markers for bone metabolism. Radiological imaging of the extremities revealed soft-tissue swelling and periostitis.
Diagnosis of PHO was made, based on mutational analysis of the HPGD gene encoding the 15-hydroxyprostaglandin dehydrogenase (15-OH-PGDH) and elevated urinary prostaglandin levels. Palmoplantar hyperhidrosis was treated with topical aluminum chloride hexahydrate solution and, due to lack of efficacy, subsequently with tap water iontophoresis. Supported by physiotherapy, antiphlogistic therapy was given to alleviate arthralgia.
Conclusion: Recently, a disrupted prostaglandin metabolism due to mutations in the prostaglandin-catabolizing enzyme 15-OH-PGDH turned out to represent the cause of PHO. Highly elevated prostaglandin levels putatively result in glandular and osteogenic stimulation and connective tissue hypertrophy leading to the mainstay triad periostosis, pachyderma and digital clubbing. In most cases, hyperhidrosis can be observed in addition. Currently, therapy is only symptomatic and mostly with limited success. However, symptoms may spontaneously ameliorate with age.

PP025 - PROLIDASE DEFICIENCY AS A CAUSE OF LEG ULCER IN CHILDREN

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Background - Prolidase deficiency (PD) is a rare, pan-ethnic, autosomal recessive disease resulting from a nonsense mutation of the prolidase gene located on chromosome 19. The deficiency of this enzyme impairs proline recycling and consequently the synthesis of collagen. This defect may be asymptomatic or associated with different clinical manifestations being the most frequently reported chronic skin ulcers.
Case Report - The authors describe a case of a 14-year-old boy with the diagnosis of prolidase deficiency since his 4, confirmed by reduced prolidase activity in erythrocytes. He was admitted in our department due to ulceration in his left foot surrounded by dry pruritic eczematous eruption. The ulcer had a polygonal form, elevated borders and a spongy base. A skin biopsy showed no specific abnormalities, except fibroblastic proliferation and inflammation. He had also a personal history of recurrent respiratory infections, learning disability, unusual facial appearance and hepatosplenomegaly. The family history was unremarkable and there was no parental consanguinity. He started treatment with 5% glycine and 5% proline ointment with significant improvement. After 8 months of follow up there was no evidence of new lesions.
Conclusion - Prolidase deficiency is a rare genodermatosis that must be considered in the differential diagnosis of leg ulcers that develop at an early age. Effective treatment is still not available but topical application of proline and glycine ointment seems to provide clinical benefit.

PP026 - MIXED CLASSICAL AND VASCULAR FEATURES OF EHLERS-DANLOS SYNDROME

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Background: Ehlers-Danlos syndrome (EDS) is a heterogeneous group of inherited connective-tissue disorders characterized by joint hypermobility, cutaneous fragility and hyperextensibility. Variants of EDS were reclassified into six major types in 1997.
Case report: A 13-year-old boy was referred to our department due to easy bruising. He was the first child of non-consanguineous, healthy parents, but born prematurely and hypotonic. Major patient features were a thin, translucent skin with visible veins and slightly extensible, elongated face with thin lips and philtrum, high-arched palate, small chin, thin nose and large bulging eyes, acrogeria, atrophic scars, joint hypermobility, flat feet, dorsal kyphosis, low muscle tone, easy bruising, previous inguinal hernia and learning disabilities. CBC, biochemical and coagulation studies, echocardiography and abdominal US were normal. Skin biopsy disclosed thin and lax dermis, without alterations of elastic fibers, but electron microscopy showed significant abnormalities in the structure of collagen fibers and fibrils of the reticular dermis. An abnormal efficiency of secretion and electrophoretic mobility of type V procollagen in cultured dermal fibroblasts were observed. However, genetic analysis did not identify mutations in COL5A1 or COL5A2 genes. The patient died at 15 years of age due to aortic dissection.
Conclusions: This case share clinical features of both classical (collagen abnormalities) and vascular (cause of death). In fact, more than 1/3 of persons with EDS do not fit exactly into a single type; overlap is common. The complete sequencing of genes COL5A1 and COL5A2 did not revealed any alteration, but other genetic alterations cannot be excluded.

PP027 - HIGH RISK SKIN CANCER IN DE SANCTIS-CACCHIONE SYNDROME: CASE REPORT

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Background: Hypersensitivity to UVB and free radicals damage in Xeroderma Pigmentogum (XP) causes a 1000-fold increased risk of skin cancers before age 20. Neurologic deterioration accompanied with XP is called "De Sanctis-Cacchione syndrome".
Objective: to report a patient of De Sanctis-Cacchione syndrome presented with a giant SCC among multiple facial SCCs.
Case Description: A 9-year-old boy, presented with rapidly growing 10x12 cm ulcerative lesion on the left cheek. He had pigmentation changes in the sun exposed skin, multiple lesions in the face and degenerated globe covered by the tumor. He was microcephalus, had psychomotor retardation, ataxia and spasticity and he was also bedridden and incontinent. The brain CT scan showed severe atrophy. Skin biopsy of the multiple lesions was SCC.
The huge tumor was excised with 1 cm safe margin, and chemoprophylaxis with oral isotretinoin was started. No recurrence was detected within 12-month follow up.
Discussion: XP is a group of rare autosomal-recessive inherited disorder of deficient DNA repair. Despite normal skin in other DNA repair disorders, SCC is unique for XP. XP is characterized by extreme skin sensitivity to UV, abnormal pigmentation and skin cancers, developed more than 50 years earlier than average. XP accompanied with neurologic deterioration is termed De Sanctis-Cacchione syndrome. Presence of progressive neurologic involvement and the age of symptom onset correlate with the degree of defect of DNA repair. Despite reports of relatively subtle dermatologic abnormality in this syndrome, it is the unique case presented high risk SCC.
Conclusion: UV-light protection and regular monitoring by a dermatologist and ophthalmologist is important in patient management.

PP028 - COSTELLO SYNDROME: A CASE REPORT

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Introduction: Costello Syndrome is a rare genetic disease associated with multiple congenital anomalies, failure to thrive and developmental delay that is due to mutations in the HRAS gene on chromosome 11p13.3.
Case report: Our patient presented at birth with respiratory distress, coarse facial features, cryptorchidy, hypoplastic scrotum and penis and severe stenosis of the pulmonar cardiac valve and failure to thrive with developmental delay.
At 12-year of age he shows curly hair with thick eyebrows

and synophridia, low implanted ears and thick lips covered by papillomatous warty lesions that were also present around nasal folds, hands, elbows and perianal area; many melanocytic nevi , slight hyperpigmentation and increased palmar and plantar creases with thickened skin in palms and soles. A cranial tomography (when 7-yr-old) showed a temporal arachnoid cyst, periventricular demyelination, periventricular leucomalacy and frontal atrophy.
He had a bone age of 1 year when he was 3-yr-old. Serial radiographic studies revealed a thin cranial vault and shortening of first metacarpal bones. Cariotype was normal (46XY). Mutation analysis using genomic DNA found a mutation (G12S) in HRAS gene in our patient but was absent in both parents; thus confirming the mutation as de novo.
Discussion: We report a further case of a patient with characteristic phenotypic and genotypic features of this rare genodermatosis. Dermatologists aware of such findings may help to early diagnosis and to avoid unnecessary diagnostic tests.
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PP029 - ERYTHROPOIETIC PROTOPORPHYRIA: A FAMILY STUDY AND REPORT OF A NOVEL MUTATION IN THE FECH GENE

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Background: Erythropoietic protoporphyria (EPP) is a genodermatosis due to impared activity of ferrochelatase (FECH), the ultimate enzyme of heme biosynthesis, resulting in the increase of protoporphyrin (PP). It causes extreme cutaneous photosensitivity and hepatobiliary disease.
Case report: A 16-year-old male presented erythema, swelling and burning sensation in the face and dorsum of the hands after sun exposure since his infancy. Symptoms worsened in summer months, with development of vesicles, exulcerations, scars in the front, nose and upper lip, hyperpigmentation and lichenification. His 18-year-old sister presented milder symptomatology, with only crusted erosions and scars in the upper lip.
Histopathology showed, in both cases, dermal deposition of PAS positive hyaline material and capillary hyperplasia. PP erythrocyte levels were > 9000 μg/L (N<1600) in both cases, but normal in parents and in a younger brother. Urinary porphyrins were normal. Rimington-Cripps test was positive.
Genetic study (GS) in both patients and mother (father's sample not available) revealed a heterozygous state for a novel mutation in FECH gene (c.1052delA) in both children.
Both children and the mother have a heterozygous low-expression single nucleotide polymorphism (SNP) IVS3-48T>C.
Conclusions: EPP is an autosomal dominant disease in most families, explained by the combined presence of a disabling FECH mutation and a common intronic SNP affecting the other allele (IVS3-48T>C), as demonstrated in our cases. The mutant allele was probably inherited from the father. Phenotypic heterogeneity for this genotype explains the divergent clinical presentation in children.

PP030 - HARLEQUIN ICHTHYOSIS SHARING FEATURES WITH NEU-LAXOVA SYNDROME

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The third child of a healthy first cousin parents was born in the 34 th week of pregnancy by ceasarean section. The skin of the newborn was covered with large, thick, white, platelike scales, partly bordered by irregular, bleeding fissures. Hands and feet were oedematous, mobility in all joints was reduced, ectropion and eclabium was remarkable. The patient's clinical features at birth were characteristic of typical harlequin ichthyosis. His brother was born with the same appearance four years before, and died at age of 2 months after an episode of sepsis.
ABCA 12 mutation was verified in homozygous fashion from DNA isolated from peripheral blood cells by PCR-RFLP method. The same mutation was detected in heterozygous fashion in his parents and his healthy sister.He was treated initially with intense emollients to skin, nasogastric feeding and acitretin 1,0 mg/kg/day from day 2. The oedema and skin tightness decraesed, and he showed white scales on his erythematous skin. His psychomotor development was severely retarded. CT scan demonstrated abnormal corpus callosum and decreased gyri.
Neu-Laxova syndrome is a lethal, autosomal recessive multiple malformation syndrome, characterized by ichthyosis, marked intrauterine growth restriction, microcephaly, central nervous system anomalies, oedema and abnormal facial features. Neonatal lethality and early death are the constant features of Neu-Laxova syndrome. The longest survival observed in an affected male was 7 weeks. Despite the absence of marked intrauterine growth retardation, survival and harlequin ichthyosis caracteristic ABCA 12 mutation, some features of Neu-Laxova syndrome are present in the reported case.

PP031 - SJÖGREN-LARSSON SYNDROME DUE TO A NOVEL MUTATION IN THE FALDH GENE: REPORT OF A CASE

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Sjögren-Larsson syndrome (SLS) is a rare autossomal recessive disorder characterized by the presence of congenital ichthyosis, spastic diplegia and mild to moderate mental retardation. It's caused by mutations in the ALDH3A2 gene, which encodes for fatty aldehyde dehydrogenase. The genotype of SLS patients varies considerably, with many private mutations in individual patients.
We report a case of a 2.5-year-old girl presented with a history of generalized scaly skin since birth. In addition, developmental milestones were delayed, she had spastic diplegia of the legs and could talk only in monosyllables. She had been born at gestational week 35, with no perinatal complications.

There was no history suggestive of a colloidion membrane. Gradually the skin lesions became thicker, with marked pruritus. Scaling and hyperlinearity of palms and soles was present.
Magnetic resonance imaging of the brain revealed unmyelinated white matter, and the proton spectroscopy demonstrated an abnormal white matter peak at 1.3 ppm.
Histopathologic examination showed orthohyperkeratosis, acanthosis and papillomatosis.
Genetic analysis of the patient detected a c.805delT(p.Tyr-269fsX5) on the exon 6 of the FALDH gene in homozygous state. The same mutation was found in heterozygous state in both parents.
The diagnosis of SLS in invariably delayed, similar to other rare diseases. On clinical grounds the diagnosis should be considered in any infant with congenital ichthyosis, especially with emerging neurological features. The ichthyosis is usually the first signal that brings the patient to medical attention, and unlike most other forms of ichthyosis, has a disturbing pruritic character. As in our case, patients tend to be born preterm. To our knowledge, the mutation of our patient was not previously reported, supporting the rich mutational heterogeneity associated with this syndrome.

PP032 - A NOVEL MUTATION IN THE KIT GENE IN AN INDIAN FAMILY WITH PIEBALDISM

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Human piebaldism is a rare autosomal dominant genetic disorder of melanocyte development characterized by a congenital white forelock and multiple depigmented patches. Previous studies have associated mutations in the KIT protooncogene with piebaldism. A Singaporean Indian family with piebaldism was screened for KIT gene mutations. We identified a novel missense mutation in exon 12 (c.1833A>C) of KIT that resulted in a leucine to phenylalanine change at codon 611. This change is predicted to affect the intracellular tyrosine kinase domain of the KIT protein, an important tyrosine kinase for the cytokine stem cell factor. This finding reinforces the association of KIT gene in human piebaldism and extends the spectrum of mutations observed in this depigmenting genodermatosis.

PP033 - A CHILD WITH TRICHOTHIODYSTROPHY AND IMMUNODEFICIENCY (ID)

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Photosensitive trichothiodystrophy (PTTD) is due to a mutation in a component of the transcription-repair factor TFIIH mostly XPD. We here present a child with PTTD due to a rare mutation in p8-TTDA : patient TTD99RO in (1).
A part form the typical features : photosensivity, collodion baby, ichthyosis, brittle hair, psychomotor delay, and growth impairement; the child has signs of hyper IgE syndrome. He has a severe atopic dermatitis needing a treatment with cyclosporin at the age of 12 years and is asthmatic. He had severe infections: a neonatal sepsis, recidivating severe respiratory and skin infections both with bacteria (S aureus, gram negatives) and fungi (C. albicans, tinea capitis due to M canis) and gastro-enteritis. Moreover he has a high arched palatum and hyper extensibility of large joints.
Blood investigations show a mild leuconeutropenia, high levels of T lymphocytes (also CD4+), IgE levels above 5000kU/L but no eosinophylia, a low response to vaccination, low IgM and IgG2 antibodies and a lowered burst activity of granulocytes.
Conclusion. Signs of ID with sometimes fatal infections have been described in PTTD (2) but they are not uniform and less well studied : leucopenia, natural killer cell defects, high IgE levels and one case with reduced CD4+ T cells (3). The signs in our patient resemble hyper IgE syndrome.
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PP034 - TUBEROUS SCLEROSIS COMPLEX- PREVALENCE DATA, CLINICAL CHARACTERISTICS IN HUNGARY AND REVIEW OF THE LITERATURE

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Tuberous Sclerosis Complex (TSC) is a rare condition inherited by autosomal dominant pattern with almost complete penetrance but variable expression. Two unrelated genes (TSC1, TSC2) are responsible for the clinical features. The lack of one of their proteins (tuberin or hamartin) leads to producing hamartomas in almost any organs. TSC may affect the skin, central nervous system, kidney, eyes, heart, blood vessels, lungs, bone, gastrointestinal tract.
The diagnosis is based on the presence of two major or one major and two minor criteries. Hypopigmented macules on the trunk and forehead fibrous plaques are the earliest skin signs in infancy. Shagreen patches, angiofibromas on the face, periungual fibromas appeare later. The neurological manifestations are present 80% of the cases. Epileptic seizures, infantile spasms, absence seizures, learning difficulties, behaviour problems, hyperactivity are most distressing features of the disease. Renal complications are the second most common cause of mortality. The lesions larger then 4 cm in diameter are associated with increased risk of serious haemorrhage. Cardiac and retinal hamartomas are usually asymptomatic, unless they are large enough or settle on critical areas.
Treatment should be symptomatic and organ specific. A multidisciplinary management approach and genetical counselling is necessary. The diagnosis often delays because the hamartomas are silent, don't cause laboratory or clinical signs for a long period or the first „harmless" skin signs (ash leaf macules) are not noticed, the general practitioner doesn't pay attention. The course and prognosis depends on the loca-

lization and size of hamartomas. The children suffering from TSC during the last five years were collected, the prevalence and clinical characteristics had been measured. The average time from the first skin signs until to set up the diagnosis was examined in Hungary.

PP035 - A KERATIN 10 GENE MUTATION IN A GREEK CHILD WITH BULLOUS CONGENITAL ICHTHYOSIFORM ERYTHRODERMA

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Bullous congenital ichthyosiform erythroderma (BCIE), also known as epidermolytic hyperkeratosis, is a generalized hyperkeratotic and blistering skin disease with mostly autosomal dominant inheritance, caused by mutations in keratin 1 or keratin 10 (KRT10). We report a case of a Greek girl with BCIE, who was found to have a mutation in KRT10 gene.

A 2-day-old girl was admitted to our hospital because she had generalized erythema, blistering and erosions since birth. Examination revealed generalized erythroderma and areas of denuded skin, without involvement of palms, soles or mucosal surfaces. Blistering improved during the first weeks of life, but was replaced by thickening and scaling. A skin biopsy taken from the lesional skin showed marked hyperkeratosis with lysis of the epidermal cells above the basal cell layer. A heterozygous C>T transition at nucleotide 466 was detected in exon 1 of KRT10 gene by direct sequencing. This nucleotide change results in an arginine to cysteine amino acid substitution at residue 156 of keratin 10 and is written K10p.Arg156Cys. This mutation was not detected in her unaffected parents, indicative of a de novo mutation. Re-examination at 2 years showed hyperkeratotic scaling of her skin with thick verrucous scale on the nape of her neck, elbow, knee and sparing of the palms and soles.

Our case confirmed that K10p.Arg156Cys mutation, which has been reported in 9 patients with BCIE from other racial or ethnic groups, is likely also to be a mutation hotspot in Greek patients with the same disease.

PP036 - CLINICAL PROFILE OF CHILDREN WITH PIGMENTARY MOSAICISM- A CASE SERIES FROM INDIA

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Backgorund- Pigmentary mosaicism results from the presence of two genetically dissimilar lines of cells which may be associated with variable abnormalities of the CNS, musculoskeletal and cardiovascular system, eye, ear and teeth.

Title- Clinical profile of children with pigmentary mosaicism- A case series from India.

Methods- All Pediatric cases (< 15 yrs), presenting with hypo/hyperpigmented bands along Blaschko lines were subjected to detailed clinical, eye, ear, skeletal and neurological examination. CT head/ MRI where possible, EEG, Echocardiography (where indicated) were performed. Data was pooled.

Observations- There were 10 children, 8 boys(80%) and 6 girls(60%) aging between 4 months to 10 yrs . Hyperpigmentation bands were seen in 6(60%), hypopigmented streaks in 3 (30%), and a combination of the two in 1 child(10%) respectively. Further categorization was as LWNH in (3), hypomelanosis of Ito (HOI) in 2, linear pigmented band in with sharp midline cut off in (2) children and remaining (3) were unclassified . Pigmentation was linear/ segmental in 5(50%), diffuse in 3 children(30%) and was confined to one side of the body in 3 children(30%) . 1 child each showed café-au lait macules and capillary malformation. The CNS symptoms appeared before 2 yrs in (80%) children. Extracutaneous abnormalities were detected in 6(60%) children and various CNS anomalies were- developmental delay (2), agenesis of corpus callosum (1), syndactyly(1), autism(1), seizures(1) and mental retardation in (1) child. Coloboma, microophthalmia, deafness, ectopic kidney, dextrocardia, syndactyly and CTEV were some of the other associated abnormalities.

Conclusion- In view of a multisystem involvement in Pigmentary mosaicism a thorough search of all the systems is recommended.

PP037 - ANOMALIES DISPOSAL: GENGIVAL FIBROMAS MOLLUSCUM PENDULUM: TUBEROUS SCLEROSIS OF BOURNEVILLE: A CLINICAL CASE

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Tuberous sclerosis (TS) is an autosomal dominant disorder characterized by focal seizures, mental retardation and skin findings, incluting congenital hypomelanotic macules and facial angiofibromas. Early analyses suggested a possible locus on chromosome 9q34(TSC1), 16p13 (TSC2). Diagnosis is essential to recognize and treat symptomatic lesions (neurological, kidney, heart, lung): causes of death in patients.

Case report. Comes to our observation of a boy 8 years, asiatic, with gingival fibromas fibroids, molluscum pendulum. Parents report the appearance of about 2 years with recurrences after surgical removal.

Consideration goal: boy is weight and height parameters and not in any event skin, with the exception of anomalies of the enamel of incisors and a macroglossia. The family history is negative for hereditary diseases. The history of remote pathological report bronchopulmonary repeated up to 6 years. The recommended diagnostic procedure requires careful examination of the goal of first-degree dove in a maternal aunt we relate eye events: amatomi syndrome and West.

At this point it is essential to the detection additional imaging célébral, videat eye, kidney and cardiac ultrasound, bone RX and genetic research.

The patient is wearing multiple asymptomatic renal cysts, and the location at a locus on chromosome 9 for encoding protein amartina.

Bone involvement, compromised vascular, neurological manifestations such as skin diseases other events are absent.

Conclusions. The diagnosis of tuberous sclerosis should be suspected when there is the presence of even one element diagnostic secondary, if two of these are present, the diagnosis is likely. The importance of placing diagnosis of tuberous sclerosis of Bourneville is crucial for the prognosis of the patient.

PP038 - EOSINOPHILIC FOLLICULITIS AS HYPERIMMUNOGLOBULIN E SYNDROME PRESENTATION IN A NEWBORN: A CASE REPORT

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Introduction. Hyperimmunoglobulin E (HIE) syndrome is a rare primary immunodeficiency, characterized by high serum levels of polyclonal immunoglobulin E with peripheral eosinophilia, recurrent staphylococcal infections of the skin and lungs, and pruritic dermatitis. Recently, STAT3 mutations have been described as a cause of HIE syndrome.
Case report. We report the case of a premature one-week-old newborn girl that presented with crops of confluent follicular pustules and vesicles on erythematous skin located on the face, scalp, ears, superior trunk and shoulders. Total white blood cell count, hepatic and kidney function tests and serum IgG, IgA, IgM and IgE were all within normal range. Cultures for bacteria, fungi and viruses were negative. The skin biopsy confirmed the diagnosis of eosinophilic pustular folliculitis. She was treated with topic erythromycin with resolution of cutaneous lesions.
In the next 4 months she was admitted several times to the Pediatric Department because of various infectious conditions, including S. aureus, Pseudomonas aeruginosa and Candida parapsilosis pneumonia with pneumatocele formation. Due to the high serum levels of immunoglobulin E (1480 UI/mL) with peripheral leukocyte count of 30800/uL with 8% eosinophils, detected at 4 months of age, the analysis of the STAT3 gene was ordered to identify the specific genetic defect in this patient with the clinical diagnosis of HIE syndrome. At 18 months of age, the patient maintains recurrent episodes of self limiting pruritic papules and pustules involving the scalp, face and trunk, becoming evident a rough appearance of facial skin.
Discussion. In children, as in adults, eosinophilic folliculitis should be viewed as a possible cutaneous sign of immunosuppression, prompting the clinician to a thorough search for an underlying disease. The prognosis of this condition depends on the opportune diagnosis and treatment of complications.

PP039 - SELECTED PRIMARY IMMUNE DEFICIENCIES WITH ABERRANT IGE PRODUCTION IN POLISH POPULATION – A CHALLENGE FOR DERMATOLOGISTS

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Background: IgE antibodies play central role in atopic diseases. However, some primary immune deficiencies (PIDs) such as hyper-IgE syndrome (HIES), Wiscott-Aldrich syndrome (WAS), Omenn syndrome (OS), immunoregulation polyendocrinopathy enteropathy X-linked (IPEX), atypical complete DiGeoge syndrome and Comel-Netherton syndrome are also associated with elevated serum IgE levels.
Objectives and Methods: In PIDs registry of Department of Immunology there are 1048 patients. Among them 7 with AD HIES with mutation in STAT3 gene, 10 with genetically confirmed WAS, 9 with OS with mutations in RAG1/RAG2 gene and 2 with Comel-Netherton syndrome.
Results: In a group of AD HIES patients newborn rash, eczema, skin cold abscesses and recurrent Staphylococcus aureus skin infections were observed. Among WAS patients eczematous lesions, multiple petechiae were ascertained in 6 patients and severe eczema herpeticum in 2 cases. All patients with OS presented with generalized erythrodermia with accompanying alopecia in 5 patients. In Comel-Netherton syndrome congenital ichtiosis, bamboo hair and eczema were reported.
Conclusions: Neonatal and infantile rashes or erythrodermias represent heterogenous conditions ranging from self-limited diseases to severe illnesses with significant mortality. If they are accompanied by systemic symptoms such as severe infections and failure to thrive, rapid diagnosis is pivotal for immediate survival and future outcome. Opportunity of performing genetic analysis in suspicion of PIDs may lead to definitive recognition of the disease and earlier institution of appropriate treatment.
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PP040 - ECTODERMAL DYSPLASIA-SKIN FRAGILITY SYNDROME: REPORT OF A CASE WITH A NOVEL MUTATION IN PLAKOPHILIN-1

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Introduction. The ectodermal dysplasia-skin fragility (EDSF) syndrome is a rare genodermatosis recently included in the hereditary epidermolysis bullosa group. It due to a plakophilin-1 deficiency, which results in desmosomal abnormality and poor intercellular cohesion among the epidermal cells.
Case report. A Caucasian 15-year-old boy was referred to our Clinic because of generalized redness and skin fragility since early infancy. The patient showed short and curly scalp hair, and the eyelashes and eyebrows were almost absent. There were scattered crusts and erosions over the whole body surface, including the face, where perioral fissuring was prominent. The nails were thickened, and the palms and soles showed diffuse hyperkeratosis and scattered fissures. A biopsy revealed widening of the intercellular spaces within the epidermis and isolated dyskeratotic cells in the upper epidermal layers. Transmision electron microscopy confirmed the desmosomal abnormality, and revealed ultrastructural hair shaft anomalies. Mutation analysis of the placophilin-1 gene demonstrated a novel mutation in exon 7, thus confirming the diagnosis of EDSF syndrome.
Discussion. EDSF syndrome is an autosomal recessive genodermatosis characterized by skin fragility, palmoplantar hyperkeratosis, onichodystrophy, perioral fissuring and noncicatricial alopecia. Several mutations in plakophilin 1 have been reported, none of them similar to that one observed in our patient(1).

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PP041 - A NEW TYPE OF COMPLEX FIBROFOLLICULAR HAMARTOMA IN TUBEROUS SCLEROSIS

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Background: We present an unusual complex hamartoma previously undescribed in a patient with tuberous sclerosis.
Case report: A boy with tuberous sclerosis had since the age of 2 a large plaque on the right side of the abdomen, which later became studded with numerous follicular comedo-like plugs. Through the years, the lesion progressed to become a large mass of keratin-filled cysts grouped on a base of thick and loose tissue. The lesion was excised and
showed a complex hamartoma with numerous epidermoid cysts and an excessive amount of collagen forming a thick fascia-like band in the dermis.
Conclusion: The uncommon skin lesion of this patient is most likely a manifestation of TS. It may be best explained as a type 2 segmental involvement, reflecting loss of heterozygosity that occurred at an early developmental stage.

PP042 - PAPULAR EPIDERMAL NEVUS WITH 'SKYLINE' BASAL CELL LAYER (PENS)

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Background: A new type of epidermal nevus is presented
Case reports: Five infants showed congenital epidermal lesions consisting of small, discrete, epidermal papules, affecting several areas of the skin surface. 2 patients had solitary lesions, whereas 3 patient had multiple small plaques (3, 7 and 11 lesions, respectively). They caused no symptoms and did not seem to show a predilection for any location. They did not follow the lines of Blaschko.
On histopathological examination, a regular acanthosis and strikingly arranged basal cells with palisading nuclei were noted.
The basal cell layer was perfectly delineated, and resembled the 'skyline' or 'eyeliner' pattern as described in Bowen's disease. Moreover, a diffusely compact orthokeratotic hyperkeratosis covered the papules. No remarkable changes were observed in the dermis.
Conclusion: We think that this may be a new type of epidermal nevus, for which we propose the acronym PENS, which accounts for Papular Epidermal Nevus with 'Skyline' basal cell layer (PENS).

PP043 - KINDLER SYNDROME. CASE PRESENTATION AND PRENATAL DIAGNOSIS

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Introduction. Kindler syndrome is an autosomal recessive genodermatosis characterized by trauma-induced blistering, poikiloderma, and photosensitivity. KS resemble EB early in the neonatal period. KS is caused by a primary defect in adhesion protein known as kindlerin, a fermitin family homologue 1.
Case report. A newborn female was seen for limited blistering at trauma sites. EB was suspected and a skin biopsy and immunofluorescence mapping were not diagnostic. She continued to develop blisters and easily burned during the summer. There was progressive poikilodermia. There was also chronic constipation. A new skin biopsy showed thickened labelling with anti type VII collagen , anti type IV collagen,and anti laminin 5 mAb and immunofluoresence labeling with anti fermitin family analoge suggested the diagnosis of Kindler syndrome.
Mutation screening of KIND1 was performed in genomic DNA by polymerase chain reaction (PCR) and direct sequencing. A nucleotide substitution (A-to-G transition) within the consensus sequence of the donor splice site of intron 11 was demonstrated at the homozygous state in the proband. The same mutation was detected in heterozygosity in the father and the mother. Prenatal diagnosis was performed in a subsequent pregnancy.
Conclusion. Kindler disease is often misdiagnosed. Molecular diagnosis and prenatal diagnosis is now possible.
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PP044 - CORRELATION BETWEEN SKIN ANOMALIES AND NEUROLOGICAL MANIFESTATIONS IN TWO SIBLINGS WITH TUBEROUS SCLEROSIS - CASE REPORT

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Tuberous sclerosis complex (TSC) is a multiorgan disorder that primarily affects the brain, skin, and kidneys. Recent advances have elucidated the genetics of this complex, which has helped lead to an increased understanding of the basic neurobiology of this disorder. There is both phenotypic and geneotypic heterogeneity. Typical cutaneous findings in tuberous sclerosis are present in over 90% of cases and represent one of the earliest markers of the syndrome.
We analyzed skin manifestations of TSC in two siblings, a boy and a girl. The severity of skin manifestation correla-

te with severity of neurologicaly manifestations. The boy presented with adenoma sebaceum, ash-leaf spots, shagreen patches, forehead plaque and severe mental retardation, epilepsy. His sister presented with ash-leaf spots, shagreen patches and epilepsy.
The treatment of epilepsy remains a major challenge in these patients. Early identification to ensure proper monitoring and genetic counseling continue to be important clinically.

PP045 - MILD PC WITH NOVEL K6A MUTATION

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Introduction. Pachyonychia congenita type I is an autosomial dominant disorder caused by mutations in keratin genes KRT6A and KRT16. Although there is usually a palmoplantar keratoderma and oral leucokeratosis, we report the case of a family presenting only few symptoms, limited to nail changes.
Observation. An 11 years old child had hypertrophic dystrophy of the nails with onycholysis and yellowish discoloration. There was no palmoplantar keratoderma or oral leucokeratosis. The mother was asymptomatic and the father could not be examined. Mutation analysis showed that the daughter was an heterozygous carrier of a mutation of the KRT6a gene, designated as p.Arg164Pro; c.491G> C, absent in his mother. Her 7 years old sister presented identical nails dystrophy without oral leucokeratosis or palmoplantar keratoderma. Although the father hasn't been examined, the pedigree suggests paternal inheritance.
Discussion. The mutation KRT6A, p.arg164Pro; c.491G> C, found in our patient, has already been reported in two families: one of them had a more severe phenotype with palmoplantar hyperkeratosis appeared between 3 and 18 years old, while the other had mild nails dystrophy. The mild features phenotype observed here is probably not due to their young age.
To explain the variation in clinical severity between families with the same mutation in KRT6A, the intervention of genetics modifier is suspected. Some other variants of keratin genes may modify the expression of the disease, but other genes such as filaggrin's may also be involved.
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PP046 - LABIAL HYPERTROPHIC HYPERKERATOSIS: NEONATAL PRESENTATION OF A WHITE SPONGE NEVUS

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Introduction. The white sponge nevus (WSN) is an hereditary disease due to mutations of keratin 4 and 13 genes. It is characterized by a whitish aspect of the mucous membranes, especially the oral mucosa which is always involved, but also nasal, oesophageal, vaginal and rectal mucosae. We report a hitherto undescribed neonatal presentation of WSN.
Case and method. A three month old girl had hyperkeratotic, hypertrophic and everted lips since birth. Oral examination showed a whitish oral mucosa. Other mucous membranes were normal. She was diagnosed with white sponge nevus, since her father had been examined thirty years earlier in our department at the age of two years with similar oral and labial features. The thickened appearance of his lips disappeared during childhood whereas the whitish aspect of the cheeks remained unchanged in adulthood. His own father and elder brother exhibited similar oral lesions. A biopsy performed in the oral mucous membrane showed parakeratosis and vacuolization of suprabasal epithelial cells.
Results and discussion. This peculiar presentation of WSN in the neonatal period has never been reported to our knowledge. The thickened hyperkeratotic lips were noticed during infancy in several family members , as clinical photographs showed. These clinical features may result from a particular KRT4 or KRT 13 gene mutation. Alternatively, this may be a common but overlooked feature of the white sponge nevus in infancy, which may have been overlooked because the diagnostic is usually made later in childhood or adulthood. In our family, lip thickening disappeared gradually.
Conclusion. As in other keratin gene disorders, neonatal presentation of the white sponge nevus can be strikingly different from its usual manifestations inlater childhood or adulthood.
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PP047 - ETIOLOGIES OF ONYCHOMYCOSIS IN SHAHRE REY, IRAN: A 5- YEAR STUDY

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Background: Onychomycosis results from invasion of the nail plate by dermatophytes, yeasts or mould species of fungi. This study shows the etiologic agents of onychomycosis in Shahre Rey, Iran.
Methods: Results of mycological tests of nail clippings or subungal scrapings collected in the period between 2002 and 2006 were analyzed. Both the direct microscopy and the cultures of the nail material were performed to identify the causative agent.
Results: Out of total of 400 patients examined, finger (15%) and toenail (85%) involvement. 40% were mycologically proven cases of onychomycosis. Among positive fungal cultures dermatophytes were isolated in 71% and yeasts in 29%. Trichophyton rubrum was the most common causative agent (52%) , followed by T. mentagrophytes var. interdigitale (27%), Candida albicans (13%), Epidermophyton Floccosum (5%), C. Krusei(3%).
Conclusion: Dermatophytes fungi, especially T.rubrum and T.mentagrophyte var. iterdigitale, were responsible for most of the infections.

PP048 - TRICHOSCOPIC PATTERNS OF CHILDRENS ALOPECIA AREATA

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Background. Trichoscopy allows to explore the hair at 10 to 800 magnifications and to observe precisely the types of hair, follicular openings,the peripilar signs and to follow up the evolution of the disease or the treatment efficacy prior to naked eye.
Objectives. To find wich are the most common trichoscopic patterns of childrens alopecia areata.
Methods. I studied 50 childrens with 89 plaques of alopecia areata by trichoscopy before and after 3 months of treatment.
Results.
1) 76% of childrens had regulary distributed yellow dots-coresponding to hyperkeratotic plugs in hair follicle
2) 46% had exclamation mark hairs
3) 44% had distrofic –broken hairs
4) 24% had cadaverised hairs-black dots in the hair follicles
5) 18 % had corkscrew hairs
6) 14 % had short pseudo regroing hairs-they are atrophic hairs and they are a sign of active alopecia areata.They mostly disappear at 3 months trichoscopic follow up.
7) 4 % had circle hairs
8) 4 % had vellus hairs-0,03 mm or less in thickness
9) 4 % had white dots-feature of fibrosis;they have extensive and two years persistent alopecia areata.
10) I didn't find any pseudomoniletrix hairs
Conclusions. The most frequent pattern of childrens alopecia areata is the presence of regular yellow dots(76 %),the second is the presence of exclamation mark hairs (46 %) The presence of pseudoregroing hairs is a sign of the activity of children alopecia areata.They are thin hairs that differs from normal thick real regroing hairs-sign of the treatment efficacy.
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PP049 - CONGENITAL MALALIGNMENT OF THE BIG TOENAIL - THE RESULT OF AN ORTHOPEDIC MALFORMATION?

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First described in 1978 by P.D. Samman as great toenail dystrophy, it was renamed by R. Baran et al. 1979 as congenital malalignment (CMAL) of the big toenail, due to another idea of its pathogenesis. We present a 4-year-old girl with typical features of CMAL of the big toenail, such as lateral deviation of the long axis of the nail, onycholysis, oystershell-like deformity and thickening of the nail plate, discoloration ot the nail, triangular nail shape as well as a sharp bend of the nail in its medial aspect. The severity of the onycholysis appears to be the most important prognostic factor.
This condition is often misdiagnosed and treated as onychomycosis. If no improvement occurs within the first two years of life, only progressive worsening has to be expected. The ideal age for surgical correction of the longitudinal axis of the nail to our understanding is at age two years. Our observations based on more than 30 cases reveal that almost all CMAL are associated with an underlying bone-anomaly of the distal phalanx of the toe. We postulate that this defect is the primary cause with the naildystrophy being the victim of this orthopedic malformation.
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PP049-bis - MONILETHRIX: A NEW FAMILY WITH THE NOVEL MUTATION IN THE KRTHB1 GENE

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Monilethrix is a dominant hereditary hair dysplasia due to a mutation in the type II hair cortex keratins: hHb1,hHb2, hHb6(1). Autosomal dominant mutations in exon-7 of KRTHB6 gene were identified in large families from Turkey and India as well as in the KRTHB1 gene(2,3).
Objective: To report a new family with the novel mutation.
Methods: A 2-years old girl presented with clinical monilethrix. Her parents, brother and sister were examinated without showing hair abnormalities.Dermoscopy and light microscopy showed periodic constriction of the
hair shaft in the girl.A genetic study was performed. Results: The molecular study showed a heterocygotic nucleotide change in the position 154 of the exon-1 of the KRTHB1 gene (154G>C)in the affected girl,her mother and her sister. No changes reported in her father and brother.
Conclusion: In the DNA sequence this substitution means a change of aminoacids at the level of the protein (substitution of glycine by arginine in the position 52) with severe structural consequences because of the substantial differences of size and lacking flexibility of the hair shaft. Gly52Arg substitution in exon-1 of the KRTHB1 gene described in our family have been reported only in a large family of Indian origin(3) and it may induce to think that this mutation is associated to an incomplete penetrance of the disease.
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PP050 - HEMANGIOMA AS THE MOST COMMON SKIN TUMORS IN INFANCY

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Background: Hemangioma usually appear shortly after birth and grow quickly during the first year. At birth they are either absent or barely evident, but they proliferate in the first few weeks to months of life, followed by an involution phase over several months to years.
Objectives: The objective of this study was to describe cha-

racteristics of infantile hemangioma and compare with infantile hemangioma referral patterns.
Methods: A child who has multiple hemangiomas in the skin should also be evaluated by a vascular anomalies specialist and pediatricians during period five years of 2004-2008. Hemangiomas have marked clinical heterogenity. RESULTS: About seven percent of children with a hemangioma develop an ulcer, typically on the lip or the peri anal region . Seventy five percent of hemangioma size was reached during the early proliferative stage at a mean age of three months. Hemangiomas are the most common soft-tissue tumors of infancy, occurring in approximately four percent of one-year-old children in Bosnia and Herezgovina.
Conclusions: Hemangiomas are the most common tumors in infancy and childhood. Referral for treatment of vascular anomalies is dependent on an accurate diagnosis of these lesions by the primary care pediatrician. More than half of hemangiomas are on the head and neck. Hemangiomas may have different appearances, depending upon the depth of the increased numbers of blood vessels.
Key words: Hemangiomas, Children, Diagnostic, Epidemiology.
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PP051 - INFANTILE HEMANGIOMA: THE SAUDI EXPERIENCE

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Background: Hemangiomas are benign localized growths of proliferating blood vessels that consist primarily of endothelial cells. Hemangiomas are the most common tumor of infancy, with an estimated incidence of up to 4% to 10% of infants at the end of the first year of life.
Materials and Methods: A prospective study enrolling new patients with infantile hemangiomas seen by a pediatric dermatologist was done.
Using a questionnaire, investigators collected information about patient gender, prematurity, maternal age, maternal illnesses, family history, age, age at onset, location, type and number of hemangioma, twin pregnancy, complications of the hemangioma, management used, duration of follow-up and evolution. Follow-up visits were scheduled according to clinical need.
Results: A total of 205 hemangiomas were observed in 120 patients. There were 98 girls (82%) and 22 boys (18%). A total of 84 (70%) of patients had lesions at birth, while 10% (12%) developed lesions within the first month of life. The majority (77.5%) of patients had a solitary hemangioma. Approximately 47 (39%) were located on the face. Ulceration was the most common complication noted in 26 patients (54.0%).
Conclusions: Our findings were similar to international findings. Lower incidence of prematurity was noted. Ulceration was the most common complication.

PP052 - FETAL ASCITES ASSOCIATED WITH FAMILIAL CONGENITAL PLAQUE-TYPE GLOMOVENOUS MALFORMATION

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Introduction: Congenital plaque-type glomovenous malformation (GVM) is the rarest form of GVM. There are familial and sporadic cases. Glomulin gene mutations have been identified in both cases. We report a newborn with fetal ascitis associated with this type of GVM.
Case report: A girl who was born preterm (36 WG) by elective cesarean due to ultrasonographic diagnosis of fetal ascites 1 week before. Examination at birth showed abdominal distension and a vascular skin lesion. A paracentesis yielded liquid of chylous characteristics. The etiological screening for ascites including lymphoscintigraphy was negative. Conservative management resulted in complete resolution of the ascites within few weeks.
Dermatologic examination at 48 hours of age showed a large red plaque with prominent vessels and cutaneous and subcutaneous atrophy on the inner left thigh and leg. There was no bruit or thrill. The girl's father had since birth several not compresible bluish nodules grouped on the left thigh. Histologic and immunohistochemical findings were suggestive of GVM in both patients.
Discussion: GVM is a malformative disease which is not usually associated with other abnormalities. The simultaneity of ascites and GVM plaque-type might be pure chance but we can not completely rule out an etiological relationship. The common mesenchymal origin of GVM and lymphatic vessels could be implicated. Fetal ascites have been anecdotally described with cutis marmorata telangiectatica congenita and with capillary malformation-arteriovenous malformation syndrome. Glomus tumors rarely involve internal organs.
Conclusions: The association of fetal ascites and GVM plaque-type has not been previously described. We emphasize the difficulty of GVM clinical diagnosis at birth and support the discussion of genetic counseling.

PP053 - BILATERAL PAROTID HEMANGIOMA: THERAPEUTIC APPROACH

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Background: Parotid hemangioma (PH) is a cervicofacial segmental hemangioma associated to potential life-threatening airway obstruction, ulceration, and risk of delay in language acquisition due to ear involvement and abnormal auditory conduction. Contrary to most infantile hemangio-

mas (IH), it frequently proliferates after the year of age and needs long-term treatment.
Case report: A 3-month-old female patient presented an extensive combined cervicofacial IH in a "beard" distribution, ulcerated on the lower lip. Craniofacial angio-MRI revealed an extensive angiomatous involvement of parotid and parapharyngeal spaces and posterior deviation of deep vascular axis of the neck and submandibular glands; however, there were no airway or intracranial compromise. Analitical, echocardiographic and ophtalmological evaluation was normal. Prednisolone (3-mg/kg/day) was started and maintained during first year of life in slow tapering, with reduction of size and tonality of the lesion, despite failure to thrive and arterial hypertension. A month after withdrawal of oral steroid (OS), growth and tension increase of the lesion were observed. Therefore, we decided to start propanolol (PN) at a dose of 2-mg/kg/day and, gradually, an evident regression, lightening and palpable softening of the tumor were disclosed, objectified by imagiologic studies. At present with 22 months old, the patient is in the 9th month of \beta-blocker therapy without noticeable side effects.
Comments: PH is an alarming tumor requiring, in most cases, systemic treatment, usually with OS. Recently, PN was reported in the treatment of proliferative and complicated IH and its beneficial effects and safety profile were confirmed in the present case.

PP054 - AN OPEN STUDY OF PROPRANOLOL FOR INFANTILE HEMANGIOMA

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Background: Propranolol has been used successfully in a limited number of children with infantile hemangiomas (IHs).
Study design: A multicenter open study describing the efficacy and adverse effects of propranolol in IH.
Methods: Seventy-one infants with IHs were treated with oral propranolol, 1 mg/kg/12 hours, for at least 14 weeks. All patients underwent a cardiologic examination and monitorizing during the period of study. A picture-based severity scoring system carried out by 5 observers was used to evaluate efficacy, considering 10 as the original IH before treatment and 0 as completely normal skin. The mean of the 5 independent measurements was used in the analysis.
Results: Propranolol was a rapid and effective treatment for IHs at 4 weeks ($p < 0.001$), at 8 weeks ($p < 0.001$ comparing with the 4 weeks value), at 12 weeks ($p < 0.05$ comparing with the 8 weeks value), and thereafter up to 32 weeks ($p < 0.01$ comparing with the 16 weeks value). The response of IHs to propranolol was similar regardless of sex, age at onset of treatment, type of involvement (segmental and non-segmental), facial segments affected, special locations (eyelid, nasal tip and parotid region), ulceration, and depth of IHs. Five infants with airway IHs causing air flow obstruction dramatically improved within a few days of propranolol therapy. Very few side effects were reported, mainly agitated sleep in 10 of 71 patients.
Conclusion: In the series of patients in this observational study, oral propranolol 2 mg/kg/day was a well-tolerated and effective treatment for IHs. Prospective studies are needed to establish the exact role of propranolol in the treatment of IHs.

PP055 - LOWER EXTREMITIES ANNULAR LESIONS IN YOUNG ADOLESCENT – PURPURA ANNULARIS TELANGIECTODES OF MAJOCCHI

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Background: Purpura annularis telangiectodes of Majocchi (PATM) is a rare form of pigmented purpuric dermatosis (PPD) of unkown etiology, thought to be secondary to capillaritis. It is characterized by symmetrical, purpuric, telangiectatic and atrophic patches, with a predilection for the lower extremities and buttocks of adolescents and young adults. Coalescence into annular patches and plaques is common, but pruritus is a rare event. Involution usually occurs in the course of a year; however, relapse is frequent. Treatment is mainly based on postural measures to decrease venous stasis, and topical steroids if itching or eczema are present.
Case report: A 16-year-old female presented with a 1-month history of pruriginous, nonblanchable and purpuric plaques and patches symmetrically distributed in both lower limbs, with 2 to 10-cm in diameter, annular configuration and central lightening. Personal and family history was irrelevant. Laboratory tests, including hemogram with platelet count, sedimentation rate, biochemistry, complement, coagulation study and immunologic panel were normal or negative. The skin biopsy showed perivascular lymphocytic dermatitis of the superficial dermis, with abundant extravasation of erythrocytes and siderophages. The diagnosis of PPD, purpura annularis telangiectodes of Majocchi type, was proposed. The benign nature of the disorder was explained, and the patient was advised to maintain leg rest, use compression stockings and apply a topical steroid cream in the pruriginous lesions. Gradually, the lesions faded out with no residual dyschromia or scar.
Comments: Despite being a benign condition, PATM is frequently misdiagnosed due to the purpuric (usually petechial) nature of the lesions, leading to the exclusion of thrombocytopenia and vasculitis.

PP056 - ARE THERE SEVERE SIDE EFFECTS OF PROPRANOLOL THERAPY IN CASES OF INFANTILE HAEMANGIOMAS?

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Background: The propranolol therapy for cases of rapidly progressing infantile haemangiomas started in 2009, the patients had been selected from the Haemangioma-Vascular Malformation Ambulance of Heim Pál Children's Hospital. The working group consists of a pediatric dermatologist, an angiologist, and an interventional radiologist expert.
Methods: The propranolol therapy had been started in 20 cases, between age of 2-10 month old. The age, localisation,

the extent of haemangioma, the progression had been taken into consideration as a selection criteria.
The status had been determined by photo documentation CT and or MRI, cardiological, ophthalmological, neurological examination, and laboratory test had been performed. The beginning the dose was 2mg/ kg/die divided in two parts. At each case the blood pressure, the pulse, the serum glucose level was measured.
Results: Two of the twenty patients propranolol therapy had to be reduced due to severe bradycardy. At each patient half dose was given, but the therapeutic effect was not adequate at the dose of 1mg/kg/die. In case of another female infant due to the haemangioma progression in the orbital region, which appeared as a severe threatening amblyopy a higher dose of propranolol was administered. The tumour became softener after two days of treatment.
While the serum glucose level is instable, regular laboratory tests were performed. At the dose of 2mg/kg/die propranolol one case showed severe bleeding. After the reduction of the dose the exulceration healed.
Conclusions: The possible side effects observed by the working group were: bradycardy, instabile serum glucose level, severe bleeding. The therapy was very effective in all cases, the side effects were mild and transient. Due to „off label" use, official permission were administered for each patient from the Ethical Comitte of the Hungarian National Pharmaceutical Institution.

PP057 - FURTHER EXPERIENCE WITH PROPRANOLOL FOR INFANTILE HEMANGIOMAS

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Infantile hemangiomas (IH) are the most common benign tumors of infancy and may be associated with significant morbidity. Dramatic improvement of complicated IH with the use of propranolol was recently reported.
Here we report further experience with propranolol for complicated IH. Ten patients aged 1.5 - 11 months with 17 hemangiomas were treated with propranolol at a dose of 2-3 mg/kg/day for 1-11 months and their treatment was followed also by color doppler ultrasound. Eight of the hemangiomas were located on the face and neck region, two of which were segmental. Four of the hemangiomas in 4 patients were ulcerated. The ulcer of one hemangioma located on the chin-neck area regressed just with propranolol, but for the other 3 ulcerated hemangiomas which were located on the chest, nose and genital area in 3 different patients addition of systemic corticosteroids was needed. With the addition of corticosteroids ulcers healed in these 3 lesions but overall clinical response in 2 of these hemangiomas were not so cosmetically acceptable, although 3 additional lesions in one of the patients regressed favorably. Prompt and cosmetically good responses were observed in the other 7 patients who used only propranolol.
No major side effects were reported except for sleep disturbance in one patient. Prompt and favorable responses observed in majority of our patients makes further confirmation of propranolol as a safe and effective treatment alternative in hemangiomas necessitating manipulation.

PP058 - A CASE OF CONGENITAL TUFTED ANGIOMA WITH SPONTANEOUS REGRESSION

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Tufted angioma is an uncommon, histologically benign vascular tumor. Lesions usually present during infancy or early childhood. They are usually reported to persist, often slowly enlarging and may be tender. Congenital forms are rarely reported.
Here we report a 3 month old boy with a congenital bluish plaque in the abdominal area. Histopathologic examination revealed lobules of capillaries in the dermis, each surrounded by dilated crescent shaped vascular channels leading to the diagnosis of tufted angioma.He was in good general health with no associated problems and the lesion regressed completely at the age of 11 months. Though spontaneous regression is rarely reported with tufted angiomas in general, congenital forms are reported to be more prone to spontaneous regression. So if there is no vital organ or functional compromise just observation without any manipulation can be offered for congenital forms of tufted angioma.

PP059 - RISK OF OCCULT DYSRRAPHISM IN CHILDREN WITH LUMBOSACRAL CAPILLARY MALFORMATION. STUDY WITH SPINAL ULTRASOUND AND MRI

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Introduction: Cutaneous lesions at the lumbosacral area can be markers of hidden spinal dysrhaphism (SD). The value of capillary malformations has been questioned. Spinal MRI is the gold standard procedure to rule out SD, but is expensive and requires sedation. Spinal ultrasound, performed before 6 months of age, could be an alternative screening method.
Methods: 35 children less than 6 months of age presenting to our clinic for lumbosacral capillary malformations from January 2001 to November 2009 were included. Spinal ultrasounds were performed in all of them. MRI was also performed, after one year of age, in 13 patients.
Results: There was another skin SD marker in 6 patients: sacral dimple in 4; hypertrichosis in 2. Medullar cone location was determined in all patients with ultrasounds. Two patients (5.7%) had abnormal ultrasounds: a fillum terminale cyst and an intraespinal lipoma repectively,that were later confirmed by MRI. The first case had an isolated capillary malformation and the second also had diffuse mild hyperthricosis. None of the other patients studied with both techniques had any abnormality.
Conclusions: Spinal ultrasound before 6 months of age can be a useful, non-invasive and non-expensive screening procedure for SD. We observed a good concordance between ultrasound and MRI.
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PP060 - EXPRESSION OF PROX-1, A LYMPHATIC ENDOTHELIAL TRANSCRIPTION FACTOR, IN KAPOSIFORM HEMANGIOENDOTHELIOMA AND TUFTED HEMANGIOMA

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Kaposiform hemangioendothelioma (KHE) and tufted angioma (TA), low grade malignancy vascular neoplasm, are rare tumors mainly occurring in early childhood. Previous studies demonstrated that podoplanin, a well-characterized lymphatic endothelial marker (recognized by D2-40 monoclonal antibody) is expressed in KHE and in TA. The expression of other sensitive and specific lymphatic markers, such as Prox1, has been investigated in vascular tumors. Although the role of Prox1 as a transcription factor regulating the lymphatic vascular phenotype is well established, its function in vascular tumorigenesis remains unexplored. Our recent results demonstrated that Prox1 expression promoted an aggressive behavior in murine models for kaposiform hemangioendothelioma. We optimized immunohistochemical staining for human Prox1 and examined its expression in 39 vascular neoplasms: KHE (n = 11), TA (n = 11), infantile hemangioma (n = 13) and pyogenic granuloma (n = 4). Prox-1 was expressed strongly and diffusely in the spindled tumors cells of 11/11 KHE (100%) and moderately but focally in the tufted nodules of 10/11 (91%) TA. All other tumors were negative. Our results demonstrates that Prox1 is a specific diagnostic marker for KHE and TA among other pediatric vascular tumors and that it may be a marker for malignant potential in vascular neoplasms.

PP061 - HAEMANGIOMA INFANTILE EXULCERANS (CASE REPORT)

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Infantile haemangiomas are benign proliferations of endothelial tissue, and are the most common tumors arising in the neonatal period. They are characterized by a significant postnatal growth during the first several months of life, followed by a slow spontaneous involution over the ensuing years. Ulceration is the most common complication occurring in up to 16% of cases.
We are presenting a 9-month-old female infant with a superficial haemangioma on her left labia majora. It started to develop at the age of 2 weeks. Within a month an ulceration appeared. Since it failed to respond to a local therapy application, systemic corticosteroids (methylprednisolone) were introduced with a starting dose of 2 mg/kg for 8 weeks. Within the following few months the dose was gradually tapered. Satisfactory therapeutic results were achieved – cessation of growth, ulceration closure, and shrinkage of the tumor. Neither side effects nor rebound phenomenon to the prescribed therapy were observed during the follow up period of 6 months.

PP062 - A STUDY OF INFANTILE HEMANGIOMAS

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Background. Infantile hemangiomas are the most common tumor of infancy.
Objectives. This study aims to identify clinical characteristics associated with complications and treatment of infantile hemangiomas.the need for therapeutic intervention.
Methods. A prospective study of 50 children with infantile hemangiomas who were below 12 years of age were taken up for the study.
Results. In our study, 66% of hemangiomas were present at birth, 22% were seen between 1-5 years of age, 10% appeared by first month of life and 2% appeared after 5 years of age. Also, it was seen that 90% of hemangiomas were of superficial type and 10% were of deep type. Regarding the number of hemangiomas, 84% of children had single hemangioma, 10% had 2-5 lesions, 4% patients had 6-10 lesions and 2% patients had more than 10 lesions. Positive family history was seen in 8% children. The commonest site of involvement was head and neck seen in 56% patients, trunk involvement was seen in 28% patients and extremeties were involved in 16% of children. The commonest complication was ulceration seen in 12% patients.
Conclusion. Because hemangiomas proliferate rapidly in the first few weeks to months of life, there may be a window of opportunity to intervene in high-risk hemangiomas, in an attempt to prevent complications, including permanent scarring.

PP063 - A CASE OF FAMILIAL GLOMANGIOMATOSIS

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We report the case of a 2 year old boy presenting with multiple asymptomatic, blue-violet vascular papules which had progressively appeared after birth. These were localised on the head, arms and lower back. Family and personal history were unremarkable except for the presence of a single similar lesion in his mother and his sister. Histopathological and immunohistochemical analysis revealed the presence of glomangiomas with focal glomangiomyomatous differentiation.
Glomus tumors are benign cutaneous neoplasms that are derived from specialized arteriovenous shunts that occur normally in many parts of the body. A classic triad of pain, pinpoint tenderness with blunt palpation, and hypersensitivity to cold are noted in digital lesions, however this is less evident in patients presenting with multiple lesions. There are 3 histological types: "sensu strictu" glomus tumor, glomangioma and glomangiomyoma; the glomangioma being the most frequent. Familial cases posess autosomal dominant inheritance and are linked to mutations in the glomulin gene.These tumors do not usually involute over time

and patients request treatment for symptomatic relief and/or cosmetic reasons. Simple surgical excision is the treatment of choice even though not always feasible for multiple lesions and should therefore be reserved for symptomatic ones. Other reported therapies include sclerotherapy, carbon dioxide and argon laser, or a combination of both as well as electron beam radiation. This familial case is one of only approximately 2 dozen reported in the literature.

PP064 - SCROTAL CUTANEOUS CHYLOUS REFLUX

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A case of cutaneous chylous reflux into the scrotal skin is reported in an eight year old boy. The patient had multiple vesicle-like lesions discharching a milk-like fluid which was pure chyle. It drained from the mesenteric lymph vessels and refluxed retrogradely through abnormal vessels to the scrotal skin.

Key words: scrotal chylous reflux,chyle,abnormal mesenteric reflux lymphatics.

The normal route of lymph flow from the bowel to the blood stream is through the cisterna chyle and the thoracic duct. The term chylous reflux is used when there is back flow of chyle into abnormal areas, usually in the lower half of the body.It may show itself clinically in many different ways (1) with or without skin involvement. The congenital variety is seen with primary lymphedema which is associated with marked abdominal incompetent lymphatic tortiousity and hyperplasia or "megalymphatics". Obstruction of these lymphatics occurs continuously or intermittantly, leading to continuous or intermittant lymhedema and retrograde lymph or chylous reflux(2). Secondary lymphedema as a result of lymphatic obstruction caused by a tumor,trauma,irradiation or filariasis can also be associated with chylous reflux(3). When the skin is involved with the chylous reflux, the condition is termed cutaneous chylous reflux. The latter condition is caused by numerous dilated lymphatics in the papillary dermis and in places might perforate the epidermis(2).

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PP065 - CHARACTERIZATION OF 2 CASES OF CHILDHOOD BULLOUS SLE: AN UNDERRECOGNIZED CLINICAL ENTITY IN CHILDREN

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Background: Bullous systemic lupus erythematosus is an uncommon subset of systemic lupus erythematosus (SLE). It is rarely found in children. At initial stage, the clinical presentations may be similar to that of chronic blistering disease and may not fulfill the American Rheumatism Association (ARA) criteria for diagnosis of SLE.

Little data exist about the characteristics of childhood bullous SLE.

Objectives: To report our experience in two pediatric patients with bullous SLE.

Patients: This study explored two cases of childhood bullous SLE in nine-year period at Srinagarind Hospital. Clinical and demographic features as well as the histopathological/immunological studies collected from medical records and electronic medical records were analyzed retrospectively with special emphasis on the clinical and the diagnostic characterizations.

Results: The lag time between onset of symptoms and the diagnosis in both cases were 2 and 5 months. The early symptoms were fever, physically debilitated and fatigue followed by a gradual onset of generalized vesiculobullous eruption. In both cases, diagnoses of chronic bullous dermatosis of childhood were given. The other interesting difference between bullous SLE and the chronic bullous diseases were the appearance of oral ulcers and anemia (hemoglobin levels of 8 and 7 g/dL). In early stage, the dermatologic manifestations were indifferent between bullous SLE and chronic bullous dermatosis of childhood.

Conclusions: We conclude that childhood bullous SLE should be recognized in children with bullous dermatoses who do not respond to the treatment of bullous disease and have significant fever, fatigue, oral ulcerations and anemia in the beginning.

PP066 - LOCALIZED SCLERODERMA (MORPHEA) IN CHILDREN: TRIPLE-THERAPY OF MTX, SYSTEMIC CORTICOSTEROIDS AND TOPICAL CALCIPOTRIOL OINTMENT – 2 CASES

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Background: Morphea, usually localized and often self-limiting, can involve wide areas of the body surface area and cause severe functional and cosmetic disabilities.

Objective: To evaluate the efficacy and safety of methotrexate (MTX) and systemic corticosteroid in combination with topical calcipotriol in children.

Methods: Two girls (5 and 11 years of age) with progressive scleroderma (non-responders to systemic antibiotic treatment, topical therapies and PUVA-bath) were treated with high-dose intravenous corticosteroids (500 mg methylprednisolone) on 3 consecutive days in weeks 1 and 2, followed by oral prednisone (0.5 mg/kg day 4-7, 11-14). After 2 weeks, 10 mg of subcutaneous MTX were introduced and continued for at least 6 months. Concomitantly, topical calcipotriol was applied once daily. Safety evaluation was done on a regular base.

Results: 6-8 weeks after the first dose of methotrexate disease stabilized with no further expansion of scleroderma, followed by regression and softening of the pre-existing indurated plaques. The improvement was seen not only in the clinical observation, but also in a reduced thickness of skin measured by 20 Hz ultrasound. After 6 months of stable disease, MTX was discontinued. One patient had recurrence of disease activity. Therefore therapy was restarted, showing

again a rapid response. No adverse event due to drug administration was seen.
Conclusion: In children, special forms of morphea, e.g. generalized manifestations or rapidly progressive types require an effective and safe therapy regimen. It has to be introduced early to prevent ongoing functional and cosmetic impairment. We recommend to combine systemic therapies such as systemic steroids and MTX with topical calcipotriol.
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PP067 - "BUTTERFLY" RASH AS ONLY SIGN OF JUVENILE AMYOPATHIC DERMATOMYOSITIS
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A 9-year-old boy was referred for evaulation of a facial rash to the dermatological outpatient clinic with suspicion of lupus erythematosus. Physical examination revealed an erythematous rash across the malar surface and slight edema and erythema of upper eyelids, but there was no clinical evidence of muscle weakness or any alterations of the internal organs. The autoantibodies were negative and the levels of muscle enzymes were normal. Neurological, ophthalmological, cardiological, chest X-ray examinations gave negative results, as well as ultrasound, EMG and MRI examinations of the muscles excluded the possibility of myositis. A biopsy specimen from the skin showed a vacuolar interface dermatitis consistent with a diagnosis of dermatomyositis. The disease was diagnosed as (suspicion of) juvenile amyopathic dermatomyositis and oral chloroquine therapy was commenced, with local tacrolimus ointment treatment and light protection. In the first weeks of the therapy, moderate improvement was observed, and 8-10 months later he was absolutely asymptomatic.

PP068 - TREATING FIBROSIS IN MORPHEA WITH WATERFILTERED INFRARED-A LIGHT
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Morphea is a chronic inflammatory disease of dermal and subcutaneous connective tissue resulting in fibrosis. Current treatment strategies, such as methotrexate, topical and systemic corticosteroids as well as systemic antibiotics aim at reducing the inflammation in order to minimize fibrotic sequelae. Their effect on established fibrosis, however, is poor. Other treatment modalities such as ultraviolet A1 phototherapy are costly and not always easily accessible. We observed clinically significant improvement of fibrosis in terms of reduction of palpable induration and increase in skin elasticity as well as reduction of hyperpigmentation and subjective improvement of lesional discomfort in areas of morphea treated with waterfiltered Infrared-A radiation (wIRA; Hydrosun 500, Hydrosun Medizintechnik GmbH, Muellheim, Germany). This treatment was applied 2-3x weekly for at least 5 weeks in three adolescents. It was well tolerated without any adverse events, patient compliance was excellent. Based on these observations and on published data (1) wIRA appears to be a promising treatment modality for morphea, particularly in the so far unmet need of improving cutaneous fibrotic sequelae.
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PP069 - CRYOTHERAPY VS. TOPICAL 10% KOH SOLUTION IN THE TREATMENT OF MOLLOSCUM CONTAGIOSUM
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Background: Molloscum contagiosum (MC) is a fairly common poxvirus infection of the skin and mucous membranes especially in children. Cryotherapy is regarded as the treatment of choice for MC, however, there are some disadvantages associated with this treatment modality. In order to overcome these disadvantages other treatment options have been recommended. The objective of this study was to compare the effectiveness of topical 10% KOH preparation with cryotherapy in the treatment of MC.
Methods: In this study 30 MC patients (15 males and 15 females) with a mean age of 6.5 years, were randomly divided into two groups. In the first group, 15 patients received 10% KOH preparation as a topical solution to be applied to their lesions twice daily until the lesions became inflamed or superficially ulcerated. In the second group, cryotherapy was performed weekly for a maximum of four weeks. After one month, the patients in both groups were evaluated for response to treatment and any possible side-effects.
Results: The results of the study showed that 13 out of 15 cases (86.6%) treated with 10% KOH had complete cure. In the group treated with cryotherapy, 14 out of 15 cases (93.3%) showed complete cure. There was no statislically significant difference between the two groups ($p>0.05$). Side-effects encountered in both groups were predominantly hyper and hypopigmentary changes which were transient in nature and improved with time.
Conclusion: From this study, it can be concluded that topical 10% KOH solution is a safe, cheap and effective treatment modality for molloscum contagiosum.

PP070 - STEVENS-JOHNSON SYNDROME ASSOCIATED WITH ATYPICAL PNEUMONIA CAUSED BY CHLAMYDIA PNEUMONIAE
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Background - Stevens-Johnson syndrome (SJS) is an uncommon, life-threatening dermatological disease that is almost always drug-induced. Infectious agents have been implicated infrequently, being Mycoplasma pneumoniae the most reported culprit.
Case report - A 6-year-old boy, otherwise healthy, was admitted for the management of a generalised maculopapular eruption, in the setting of an acute episode of productive cough, headache, fever and purulent rhinorrhea. The rash had a purpuric hue, atypical target lesions and some bullous and eroded areas. There was also an extensive involvement

of the conjunctiva and oral mucosa. At the admission chest X-ray revealed interstitial pulmonary infiltrate. Laboratory evaluation showed leukocytosis, neutrophilia and increased CRP concentration. Multiple serological testing, sputum and blood cultures for the detection of bacterial and viral agents, such as Mycoplasma pneumoniae, Chlamydia pneumoniae, HSV 1 and 2, EBV, CMV, hepatitis A, B and C, were all negative. Histological examination of the skin supported the diagnosis of SJS. There was no history of drug intake in the previous 3 months. The outcome was favourable within several days, following the administration of intravenous immunoglobulin (IVIG 0,5g/kg/day,4days), clarithromycin and supportive measures. The patient was discharged 25days after the admission, with no apparent sequelae, even after 6 months of follow-up. All laboratory values gradually returned to normal during one month after discharge. However, an IgG antibody for Chlamydia pneumoniae was found positive at this time.

Conclusion - This case illustrates a pediatric SJS associated with atypical lung infection caused by Chlamydia pneumoniae, which improved with antibiotic and IVIG therapies. It also supports the view that Chlamydia pneumoniae infection is a possible trigger of SJS.

PP071 - TINEA INCOGNITO

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Introduction: Dermatophyte infections of the face can show different clinical features, due to various species of dermatophytes as well as inappropriate use of topical or systemic steroids. Because of that, tinea faciei is an often misdiagnosed clinical entity, mimicking seborrhoeic dermatitis, contact dermatitis, polymorphous light eruption, lupus erythematosus and rosacea.

Case report: We report the case of a 16-year old girl having the erythema with clearly defined border and prominent pustules and papules on the face. One year ago, the patient was treated with systemic corticosteroids because of Dg: Sclerosis multiplex. After the erythema appeared on her face, she was treated for six months with topical corticosteroids, with a progressive exacerbation of the eruption.

Direct microscopic examination of the face revealed hyphae and small-spore. Cultural examination diagnosed Microsporum rubrum species.

Therapy: After 6 weeks of topical application of Klotrimazol we achieved good results.

Discusion: The term tinea incognito has been used to describe dermatophyte infections modified by corticosteroid treatment.

PP072 - DISSEMINATED MOLLUSCUM CONTAGIOSUM IN A PATIENT WITH IDIOPATHIC CD4+ LYMPHOCYTOPENIA

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Idiopathic CD4+ lymphocytopenia is rare immunodeficiency of unknown cause. As a definition, patients have depressed numbers of circulating CD4+ lymphocytes (less than 300 / microliter or less than 20 % of total T cells) with no laboratory evidence of infection with human HIV-1 or HIV-2 and the absence of any defined immunodeficiency or therapy associated with depressed levels of CD4+ T cells. Clinical spectrum may range from asymptomatic laboratory abnormality to life threatening complications. Patients may experience opportunistic infections, autoimmune diseases and hematologic malignancies. We describe an 8 year-old mentally retarded boy with giant molluscum contagiosum disseminated over the scalp, face, neck and chest areas. Because of a past medical history of recurrent skin abscesses immunologic investigations were done. Immunologic analysis disclosed low levels of IgG, IgM, low numbers of CD4+ lymphocytes and defective lymphocyte proliferation. HIV serology was negative. He was diagnosed as idiopathic CD4+ lymphocytopenia and was treated with intravenous immunoglobulin infusions and prophylactic trimethoprim-sulfomethaxasole for immunodeficiency.

For the treatment of molluscum systemic interferon alpha, topical cantharidin and salicylic acid/ lactic acid combination were used with no improvement . Lesions recurred shortly after electrofulguration. With the use of cimetidine and podophyllotoxin topically a partial response could be elicited. Molluscum contagiosum is an important pathogen in pediatric and adult CD4+ lymphocytopenia both in idiopathic and HIV associated forms. In pediatric patients with severe or unusual viral or opportunisitic infections immunologic investigations should be undertaken.

PP073 - DISSEMINATED ZOSTER IN A HEALTHY CHILD

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Herpes zoster is a localized disease caused by the reactivation of varicella zoster virus and characterized by unilateral radicular pain and a vesicular eruption that is generally limited to the dermatome. Herpes zoster is rare in childhood and especially encountered in immunosuppressed children. However, it can be seen in children without immunosuppression. Cutaneous dissemination may occur in immunocompromised persons. Disseminated zoster requires systemic antiviral therapy. Here we present a 7 year-old immunocompetent girl with disseminated herpes zoster. She had no history of varicella infection or vaccination. The dermatomal lesions were severe and bullous in addition to being painful. disseminated lesions on the face and trunk region outside the affected dermatome were noted. She also had accompanying fever and malaise. She was terated with intravenous acyclovir with regression of fever and malaise within 2 days and with resolution of lesions within 10 days. Although zoster confined to a dermatome with mild symptoms can be occasionally seen in healthy children disseminated and severe zoster in a healthy child has not been reported previously.

PP074 - PURPURA FULMINANS SECONDARY TO VARICELLA INFECTION

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It has long been recognized that varicella infection is sometimes severe and life-threatening. We report a case of purpura fulminans (PF) following varicella infection in an otherwise healthy child and discuss its diagnosis, management and prognosis.
A 6 year- old female presented to our hospital on the 7th day of varicella because of severe limb pain and ecchymotic patches in both her limbs. She was treated with fresh frozen plasma and platelets because of clotting derangement. Protein C (PC) levels were normal but free protein S (PS) was very low. Anti-PS antibodies were detected in high levels while the anti-thrombin levels were normal and the dilute Russell viper venom test and the anti cardiolipin autoantibodies were negative. Assays for factor VIII, XII and activated PC resistance were normal and common thrombotic mutations were negative. Diagnosis of PF due to an acquired PS deficiency secondary to varicella infection was made and the child started on plasmapheresis and IV heparin infusion but due to the development of a compartimental syndrome required fasciotomies of both lower limbs. She remained on daily plasmapheresis for 7 days until PS levels were stable. Blood cultures and wound swabs were always negative and improvement in her legs was noticed on daily basis. Since discharge she received split-thickness skin grafts and her PS levels have been stable.
Although exceptionally rare, PF is one of the most feared complications of varicella and can be of grave consequence. Intensive care management and timely surgery can be successful in preventing life-long morbidity and were, in this case, essential in altering the prognosis of such a spectacular disorder.

PP075 - THE EVER-WIDENING SPECTRUM OF KALA-AZAR

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Background: Kala-Azar/visceral leishmaniasis (VL) is a systemic infection caused by Leishmania species which have visceral tropism and are endemic in many parts of the world, including Portugal. Simultaneous visceral and cutaneous leishmaniasis (CL) infections are an exceptional event and have mostly been described in HIV infected patients.
Objectives/Methods: We report a case of simultaneous presentation of VL and CL in an otherwise healthy child.
Results: A 17 month male child presented to our Hospital with a history of persistent fever, vomits and a petechial rash. Parents also reported on-going loss of appetite and loss of weight. On examination he was pale, a petechial rash was present in the abdomen and limbs and an enlarged spleen was palpable. An erythematous plaque with well defined borders was present for the last 2 months on his left frontal region. Laboratory tests revealed anemia, thrombocytopenia and a raised CRP with a blood smear compatible with bacterial infection. His echocardiogram, chest X-ray and urinalysis were normal.A bone marrow aspirate and scrapping of the cutaneous lesion both revealed leishmania parasites on direct examination. Leishmania infantum was identified by PCR on the bone marrow aspirate. He was treated successfully with Meglumine antimoniate and 1 month later all clinical and laboratory features were normal and the cutaneous lesion had healed. His HIV test was negative.
Conclusion: Simultaneous VL and CL is rarely reported in the literature and is certainly a rare event in an immunocompetent host. Our report confirms that in endemic VL areas, L. infantum can also be responsible by CL and that these simultaneous infections are not exclusive of HIV infected patients.

PP076 - STAPHYLOCOCCAL SCALDED SKIN SYNDROME

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Introduction: Staphylococcal scalded skin syndrome (SSSS) is caused by strains of Staphylococcus aureus producing epidermolitic toxins which induce detachment of the superficial layers of epidermis. The mortality rate is associated with the extent of the affected skin and is up to 4%.Mucous membranes are not assaulted.
Objectives: To describe the case of a 3,5 years old girl infected with SSSS
Results: One week preceding the onset of the disease, the child developed 3 skin lesions in the form of impetigo on the perioral area. No drug was administered. SSSS presented with fever (until 38,40 C),malaise, irritability, conjunctivitis and generalized painful erythema of the trunk, face, upper and lower extremities. Within 24 hours, bullae and bubbles were formed on the trunk, extremities and face. On several spots exfoliation and revelation of a red dermal area were observed, whereas on other areas scabbing followed. On the perioral area radial fissures appeared. Nonetheless, there were no lesions on the oral mucosa. White blood cells were 12.500 with a neutrophil percentage of 70%. CRP was 1,5 (normal range until 0,5). In both the blood cultures no bacteria were developed. In the nasal secretion culture staphylococcus aureus had been isolated.
The child was treated with clindamycin for 10 days. The fever subsided the second day of treatment and the skin lesions started improving the fifth and completely disappeared the ninth day.
Conclusions: SSSS generally has a good prognosis when diagnosed in time and properly treated with antistaphylococcal agents. In case of extended skin involvement, treatment should be administered in hospital.
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PP077 - AN OPEN STUDY OF ORAL IVERMECTIN FOR THE TREATMENT OF PEDICULOSIS CAPITIS

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Introduction: Head lice infestation is a common and growing

problem that primarily affects school-aged children. Pediculosis capitis produces not only pruritus but also is a common cause of school exclusion. Treatment failures due to pediculocide resistance or incorrect treatment application are common. Ivermectin is a semisynthetic drug derived from the Steptomyces avermitilis that is commonly used to treat ectoparasitoses.
Material and methods: An open study of treatment of Pediculosis capitis in children that had previously failed to at least one previously approved topical treatment. An oral aqueous solution of 0,6% ivermectin was given at a dosage of 1 drop/kg, equivalent to 200 mcgr/kg. A second dose was given 8 days after the first one. Lice eradication was verified by combing after the second dose. Follow-up ranged from 1 to 5 months.
Results: Nineteen patients, 3 to 25 year-old, over 15 kg of weight were treated. The response rate was 100%. Dead lice could be seen on the pillows by some patients and parents. Nits were still observed after treatment. There were no side effects from treatment. Six patients had a recurrence of head lice 2 to 4 months after treatment and were retreated.
Discussion: In our experience oral ivermectin is a very effective and convenient treatment for head lice. Cure rates were higher than those for topical pediculocides used in Spain. Incorrect treatment as it occurs with topical treatments is unlikely to occur with oral ivermectin.
Conclusions: Ivermectin in an aqueous solution at dosages of 200 mcgr/kg is an effective, inexpensive, safe and easy to use treatment for head lice.

PP078 - CONCOMITANT VIRAL EXANTEMS IN CHILDREN - DIAGNOSTIC CHALLENGE

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Background: Exanthems in children are extremly common and their expression ranges from nonspecific rashes to erruptions with distinct distribution.
New opinions of infections, the vactinations and associated changes of viral antigenic qualities influence clinical symptomatology. Viral exanthems can present a diagnostic challenge for clinicians
Objective: This study aims to detect the concomitant viral exanthems in childhood and the assotiation between etiological agences.
Methods: Since June 2006 to September 2009 295 children have been investigated because of unknown exantems were suspected of. According complete history, physical examination specific viral serologic examinations (ELISA, Western- Blott, PCR), viral exantems were detected.
Results: We distinguished Parvovirus B19 in 45 children , Coxackie B3, B4, B6 in 25 children and 20 children suffered from concomitant exantems - two or more etiological agences were detected in maculopapular, papulolovesicular rashes, the most extensive and hemoragical exantems were typical in this groupe of affected children. M. pneumoniae and Parvovirus B 19 together were the most frequent in laboratory findings.
Conclusion: Last years newer exantems can appear in childhood. In origin of new viral exantems offen participate too or more different types of viral agences that make clinical picture and duration off diseases different and more severe.

PP079 - NODULAR LESION OF THE TRUNK IN A SEVEN MONTH OLD INFANT

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Clinical Observation: A seven months old girl was referred to our department for evaluation of a nodular lesion of the trunk. Her history did not reveal any disorders.The lesion, which was totally asymptomatic, had been first recognized four months previously and its size increased progressively since. Examination revealed a yellow brown nodule measuring 30 by 28 mm with slight elevation, irregular lobulated outlines but no inflammatory signs, swelling or ulceration. Mucous membranes were not affected and regional lymph nodes were not enlarged. Ultrasonography was compatible with the diagnosis of lipoma. An excisional biopsy was performed under local anesthesia. What are your diagnosis?
Discussion: - Evoked diagnosis: mastocytoma, xanthoma, Spitz naevus, histiocytosis, malignant tumor (sarcoma...).
- Histopathologically, the specimen consisted of well-differentiated, foamy mononuclear histiocytes with occasional Touton giant cells leading to the diagnosis of juvenile xanthogranuloma.
Should any additional technique be performed to assess the histological diagnosis? What is the systematic work-up for patients with juvenile xanthogranuloma? Which treatment should be pursued in our case? What is the prognosis?
- The aim of our observation is to review on and discuss, according to recent data from the literature, the different nosological, clinical, histological, evolutive and therapeutic aspects of this rare condition?

PP080 - INTERIM RESULTS OF A LONG-TERM SAFETY AND TOLERABILITY STUDY OF ETANERCEPT TREATMENT IN CHILDREN AND ADOLESCENTS AGE 8 TO 17 YEARS WITH PLAQUE PSORIASIS

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Background: Published case reports of biologic use in children show promising results, but few clinical studies have assessed etanercept (ETN) in children with psoriasis (PsO).
Objectives: To evaluate safety and tolerability of long-term administration of ETN in children ≥8 years (yrs) of age with psoriasis.
Methods: This was an open-label, multicenter extension trial in subjects with PsO. In an initial double-blind placebo-controlled study, 211 children and adolescents received once-weekly ETN SC 0.8 mg/kg to a maximum of 50 mg or placebo ×36 weeks followed by a 12-week withdrawal-retreatment period. Topical standard therapy including steroids was allowed. Subjects completing the initial study were eligible for the 264-week extension trial. Safety results from a 96-week (144 total weeks on therapy) interim analysis in children age ≥8 yrs are presented.
Results: 182 subjects age 4 through 17 yrs enrolled in the extension study, 181 received ≥1 dose of ETN, and 140 (76.9%) completed week 96. Of these, 167 were ≥8 yrs old

at enrollment in the parent trial, 52% were male, 76% were white, and mean age was 13 yrs. The most common adverse events among children ≥8 yrs old were 63 events of upper respiratory infection (19.2/100 pt-yr), 48 of nasopharyngitis (14.2/100 pt-yr), and 27 of headache (8.3/100 pt-yr). Sixteen (5.0/100 pt-yr) skin papillomas were reported. A total of 285 (87.2/pt-yr) infections were reported. There were 5 (1.5/100 pt-yr) serious adverse events and no reported serious infections or malignancies.
Conclusions: In this study, open-label ETN was associated with no new safety signals at 144 total treatment weeks in children and adolescents age ≥8 yrs with severe plaque PsO.

PP081 - WELLS' SYNDROME ASSOCIATED WITH ALLERGIC ASTHMA EXACERBATION

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Backgroud - Wells syndrome (WS) is an inflammatory eosinophilic dermatosis of unknown pathogenesis characterized by clinical polymorphism, suggestive but nonspecific histopathologic findings, usually with a recurrent course and inconstant response to therapy. It's a rare disease, especially in childhood.
Case Report - A 2-year-old boy with history personal of allergic asthma presented with acute onset of erythematous plaques, distributed over lower extremities, 1 week after an allergic asthma exacerbation.
These lesions were pruritic but not painful and the patient was not febrile. His parents denied history of insect bites, recent immunizations, medications, trauma and other previous acute illness.
Laboratory evaluation revealed mild elevation of CRP and leukocytosis, with peripheral eosinophilia of 22% and increased IgE. Serological testing, blood and stool cultures for the detection of bacterial, parasitic and viral agents were negative. Histological examination supported the clinical diagnosis of WS.
The lesions completely remitted after 5days of sistemic steroid therapy. Within 6 months of follow up the lesions recurred 3 times, allways after allergic asthma exacerbation.
The respiratory symptoms and cutaneous lesions remitted with short course of steroid therapy.
The clinical and laboratory evaluation performed since the first episode was not helpful in the identification of another precipitant factor beside exacerbation of allergic asthma.
Conclusion - WS appears to be an unspecific hypersensitivity reaction in response to various exogenous and endogenous stimuli such as insect bites, infections, drug eruption or underlying internal disorders. The authors discuss the probability of a common pathogenesis and the role of IL-5 in allergic asthma and WS. To our best knowledge this is the first case where this association is reported.

PP082 - SCARLATINA IN THE 18TH CENTURY: DESCRIPTIONS-THERAPEUTIC APPROACHES

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Methods: Material was obtained through review of related with the era bibliography.
Results:For the correlative italic term "scarlatina",the term "purpurism" was initially used by doctor P.Ipitis at 1816 characterizing the disease as "exanthematic infective passion".Instead of the term "purpurism" the term "scarlatina" was later dominated and this term term which was used by I. Vouros at 1843 in medical journal "The New Asclepius", comparing the rash with the shell's red colour (scarlatto in italian-scarlet in English: red colour).In the journal Hermes the Scolar (1821) it is published that German Professor C.Hufeland (1762-1836) administrates Atropa Belladona to many patients as precautionary for diseases drug, an opinion that is also recorded in posterior publishings:the medical journal Medical Bee (1857),the first Pediatric book in greek (1871). Belladona tincture is administrated, in a dose of 2-3 drops,twice a day,in an one year's old child and gradually the doses increase.Later,in 1884,in the Practical Manual of children's diseases, page 40, the precautionary against scarlatina action of Belladona and the lately prized fenic acid,is doubted.
Conclusions: Scarlatina is memorized by many doctors-writers of the 18th century,as expected,regarding its high incidence in children's mortality at the time.
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PP083 - PERSISTANT URTICARIA IN HOSPITALIZED CHILDREN

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Background: Urticaria in most cases is acute, lasts for short periods of time and responds to usual antihistamine treatment.In some cases, though it persists, it lasts longer or easily relapses, responds poorely to usual treatment and affects importantly the quality of children's and their families' life.
Objectives:108 children with acute urticaria were hospitalized in our hospital during 2007-2008.Out of all children,11 boys and 8 girls(middle age:5 years) manifested with persistant urticaria and were our study's object.
Methods: All the study's patients unterwent thorough history taking and physical examination, hematological,thyroid and serological testing,administration of antihistamines in high doses, combination of antihistamines or systematic,in some cases,corticosteroids.
Results: Causes of persistant urticaria, in order of frequency,were :viral infections of upper respiratory (42,1

%),drugs(NSAID,antibiotics)21%, natural stimulants (hot, cold, exercise) 15,8%,auto-immune diseases (thyroiditis) 5,3%,urticarial angiitis 5,3%.
Conclusions: Viral infections of upper respiratory are the most common causes of recurrent urticaria in children due to their sensitivity to viruses.In a few cases, the causes of the disease remain unknown despite the thorough laboratory testing and cooperation between pediatricians and a specialist is necessary.
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PP084 - CONGENITAL LEUKAEMIA CUTIS. TWO CASES PRESENTING WITH BLUEBERRY MUFFIN SYNDROME

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Introduction: Leukaemia in the neonatal period is a rare event. Cutaneous leukaemic infiltration occurs in 25-30% of cases, usually as a sign of widespread disease, but sometimes can be the presenting sign. We hereby report 2 cases of congenital leukaemia (CL), where the first clinical picture was a Blueberry Muffin Baby.
Case 1: A term female infant presented a blue-red nodule in the perianal area and a large blue one on abdomen. Progressively, some small blue nodules appeared on the trunk, limbs and scalp.The rest of physical exploration was unremarkable. Initial blood count was normal. Histopathological study showed a diffuse dermal infiltrate of mononuclear cells with large hyperchromatic nuclei and an immunoperoxidase stain pattern characteristic of myeloid leukaemia. Bone marrow aspirated showed 100% of blastic cells. Cytogenetic sudies revealed the translocations t(11:19), (p21;q23) with rearrangement of MLL gen in 83% of the nuclei. The patient was diagnosed of Myeloid Leukaemia LM5. Treatment with standard chemotherapy was started. Partial remission was obtained and the skin nodules disappeared. Bone marrow transplant is pending to be done.
Case 2: A 2 month-old male presented with multiple blue nodules on the skin. Cutaneous biopsy confirmed as in the previous case the diagnosis of CL LM5.
The patient was treated with the same chemotherapic protocol and posterior authologous bone marrow transplant. Complete remission was obtained, and maintained 13 years later.
Comment. It is important to include the diagnosis of CL in a newborn with a Blueberry Muffin Syndrome. In general, the bad prognosis of CL, is even worse when the first signs appear on the skin, but we emphasize the importance of a precocious diagnosis and treatment because some cases can achieve a complete remission as in our second patient.

PP085 - RABSON-MENDENHALL SYNDROME

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Introduction: Rabson-Mendenhall syndrome (RMS) is a rare autosomal recessive insulin resistant disorder. It was characterized in 1955 by Rabson and Mendenhall; they described three patients who died in childhood because of complications of diabetic cetoacidosis.
Case report: A 12-year-old boy was referred to our department for a brownish-black velvety plaque on his neck since two years. Physical examination also revealed a short-statured boy with dysmorfic face, dental dysplasia, sexual precocity and psychomotor retardation. The histological examination confirmed acanthosis nigricans. The endocrinology study revealed mild insulin-resistance. RMS was diagnosed.
Discussion: In the pediatric population acanthosis nigricans may be associated with rare syndromic disorders, one of this is RMS. RMS is an inherited disorder caused by mutations in gene for insulin receptors and it is characterized by insulin-resistant diabetes in early childhood. Most of the patients develop severe and intractable ketoacidosis in a short time in spite of treatment.
Conclusion: We present an asymptomatic patient with slight laboratory alterations for glucose metabolism. A mild mutation that leaved residual insulin-binding activity could explain the slight severity and good prognosis of our patient.

PP086 - RECURRENT APHTHOUS STOMATITIS IN AN 8 YEAR OLD BOY SUCCESSFULLY TREATED WITH DAPSONE

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Background: Recurrent aphthous stomatitis (RAS) is characterised by intermittent episodes of multiple, usually very painful oral aphthae. Underlying diseases such as anaemia or cyclic neutropenia are found to be the cause in some cases. Differential diagnosis comprises a great variety of diseases like Crohn's disease, Reiter's syndrome, autoimmune bullous dermatoses, Behcet disease or viral infections. Topical therapy is sufficient in most cases but in severe cases systemic therapy (e.g. steroids, colchicine, thalidomide, TNF-α-blockers or dapsone) is recommended.
Case report: An eight year old boy of Austrian origin presented with a 4 year history of RAS. The family history was negative concerning RAS. The patient's history was negative for any food hypersensitivity. Physical examination revealed aphthae on the floor of the mouth, buccal mucosa, lower lip and soft palate. No skin lesions were present. Complete blood count, liver and kidney parameters, vitamin levels, zinc and, iron parameters as well as antinuclear antibodies were within normal range. Viral and mycological investigations were negative. No signs or symptoms of gastrointestinal disease were reported. Previous treatment included topical therapy with antiseptics, anaesthetics, and NSAIDs with poor effect. Systemic steroids showed no improvement of clinical symptoms.
Because of the severe course of disease and the phonetic problems therapy with dapsone 25mg per day for 4 weeks was initiated, which resulted in minor improvement of the aphthae. Therefore the dosage was increased to 50mg per day. Within 6 weeks complete heeling was achieved.
Conclusion: So far dapsone has not been reported as a treatment option for RAS in children. Dapsone therapy was well

tolerated in our patient and led to a complete healing of the aphtous lesions.

PP087 - BULLOUS DERMATOSIS IN A 4-MONTH-CHILD

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Introduction: Linear IgA dermatosis (LAD) of childhood is a rare, acquired, subepidermal blistering disease.
Case report: A 4-month-old child presented with pruriginous vesiculobullous lesions that started in the soles and generalized with involvement of the trunk, limbs and face. There was no mucosal involvement. The child was otherwise healthy, with no drug intake history or family history of a blistering disease.
Biopsy of a lesion showed a subepidermal blister with infiltration of eosinophils in the dermis. Direct immunofluorescent showed a linear deposition of IgA, IgG and C3 in the basement membrane zone (BMZ). Indirect immunofluorescent was negative.
With the hypothesis of LAD of childhood and due to the extension of the disease, the treatment consisted in dapsone combined with systemic corticosteroids.
After 3 weeks of therapy the patient presented with a hydroelectrolyte imbalance and sepsis. This serious complication caused an interruption of the treatment with a rapid relapse of the cutaneous lesions.
After recovery of this episode, dapsone and systemic corticosteroids were re-instituted, with a total duration of 21 months. The resolution of the cutaneous lesions was slow but complete. At present, the child is almost 3 years and is healthy with no relapses.
Discussion: The authors approach the difficulty in establishing the differential diagnosis of LAD with other bullous dermatosis of childhood, particularly in this case with bullous pemphigoid. It is also discussed the fundamental support of immunofluorescence, and the relationship of additional deposits of Ig (beyond IgA) in the BMZ and a resistance to treatment.

PP088 - DERMATITIS VERNALIS AURIUM

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Background: Dermatitis vernalis aurium (juvenile spring eruption) appears to be a localized variant of polymorphic light eruption affecting primarily boys aged 5-12 years and may occur in epidemics during the early spring. It is intermittent, pruritic, papulovesicular eruption. The most typical site is the light-exposed helix of the ear.
Objectives: The condition results from a hypersensitivity to UVA and (to a lesser extent) UVB, in combination with cold. There is a familial tendency to develop this condition. The eruption is self-limited and resolve spontaneously over a period of days, but tends to recur each spring for a few years.
Methods: We present a 6 years old boy with erythematous, pruritic grouped papules and vesicles developed on the edematous helices of the ears. He was playing outdoors for many hours in the previous first sunny days. He has complained at itching and burning of his earlobes. Histopathology revealed dense lymphocytic perivascular infiltrate.
Results: After restriction of light exposure, and with moderate topical corticosteroid the acute reaction was alleviated in a few days. Afterwards it was recommended to his mother to cover up his ears with appropriate clothing and to apply high-protection, broad-spectrum sunscreens.

PP089 - EPIDERMOLYSIS BULLOSA SIMPLEX WITH MIGRATORY CIRCINATE ERYTHEMA

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Background: Epidermolysis bullosa simplex (EBS) is an autosomal dominant inherited skin mechano-bullous disorder characterized by intraepidermal blistering in response to minor trauma or friction. It is caused by mutations in the genes KRT5 or KRT14 coding for the basal epidermal keratins 5 and 14, respectively.
Case Report: The authors report the case of a 4-year-old girl with a skin blistering eruption that started at her third day of life during phototherapy for neonatal jaundice. Since them she has multiple erythematous, annularly spreading, pruriginous vesicles on the trunk and extremities, mainly in areas of trauma, which exacerbate in summer. Mucous membranes and nails are not affected. The blisters heal without scars or milia as the new blisters annularly spread, but brown pigmentation appears in the central clearing lesions. The parents are not relatives and there are no other family members affected by the same skin condition, including her brother. Histological examination and electron microscopy of a skin blister corroborated the clinical diagnosis of EBS. Genetic analysis revealed a single deletion mutation (1649delG) in exon 9 of the KRT5 gene.
Conclusion: To the best of our knowledge this is the third time that the mutation mentioned above is reported. It had been associated with two different phenotypes, EBS with migratory circinate erythema and EBS with mottled pigmentation that show similarities, particularly hyperpigmentation patches after blistering. Our case, which is similar to the former, reinforces the idea that this mutation is probably associated with specific phenotypes.

PP090 - PEMPHIGUS VULGARIS AND PERSISTENT STOMATITIS

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Pemphigus vulgaris is an autoimmune, intraepithelial, blistering disease affecting the skin and mucous membranes and it is potentially fatal without treatment. Patients genetically susceptible produce autoantibodies that destroy desmoglein 3 (anti-D3), leading to acantholysis. Blister hystopathology and immunofluorescense establish the diagnosis. Mean age of onset is approximately 50-60 years, being a rare entity in children.

In Portugal there is no other paediatric case report in literature.
The authors present a 3-year-old boy with progressive mucocutaneous blisters resistant to symptomatic treatment. Infectious and drug-induced causes were excluded. Performed oral and skin biopsies demonstrated suprabasal acantholysis with intracellular IgG and C3. Anti-D3 was positive in the serum and the HLA study showed DRB1*0402 and DQB1*0503. Other autoimmune diseases were excluded. Besides progressively higher doses of systemic prednisolone the lesions persisted and an adjuvant therapy with IVIG and azathioprine was started. There was a positive outcome, but the complete resolution of the lesions was never achieved and introduction of rituximab is now being considered.
All these drugs have heavy iatrogenic effects especially in children. Therefore, the biggest challenge is to accomplish the balance between disease control and life quality.
Key words: Pemphigus vulgaris, desmoglein 3, autoantibodies, acantholysis

PP091 - A CASE OF PEMPHIGUS HERPETIFORMIS IN A 12-YEAR-OLD MALE

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Introduction: Pemphigus herpetiformis (PH) is one of the less common forms of pemphigus. PH in children is unreported. We describe a case of child which developed PH.
Observation: A 12 year old boy was seen at our department with erythematous plaques, vesicles and crusted lesions associated with severe itching persisting for six month. The boy's family history and personal history were unremarkable, and his growth and development had proceeded normally. On examination, we observed a polymorphous rash composed of oedematous papules, 3 to 4 mm in size and of a pinkish red color, arranged in herpetiform pattern. Also seen were vesicular, bullous lesions up to 0.5 mm in size, some on erythematous skin and some on apparently healthy skin without eruption, some having clear contents and some being purulent. They were seen especially over the back, buttocks, chest, abdomen, legs and arms. Mucosa and nails were normal. Nikolsky sign was negative. Histologic examination of one of the lesions showed a subcorneal bulla containing acantholytic epidermal cells. The lower epidermis showed spongiosis and focal acanthosis. Superticial and reticular dermis had an inflammatory infiltrate of eosinophile and neutrophile around some of blood vessels. Direct immunofluorescence demonstrated intercellular IgG, and C3. Diagnosis of PH was made and treatment with Dapsone 2 mg / kg mg per day resulted in total clinical remission. However, two months later, new vesicles reappeared and treatment was begun with prednisone at a dose of 2 mg / kg daily. There was a very good response; the rash regressed slowly, until it completely disappeared after 4 weeks of treatment.
Discussion: We did not found reports of pemphigus herpetiformis in childhood other than our patient. Childhood pemphigus herpetiformis is a rare disease, often initially misdiagnosed. The disease must not be forgotten as a possible cause of erosive mucocutaneous disease in children.

PP092 - LINEAR IGA BULLOUS DERMATOSIS OF CHILDHOOD – A THERAPEUTIC CHALLENGE

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Linear IgA bullous dermatosis of childhood is a rare bullous disease characterized by subepidermal bullae and linear IgA deposition on BMZ.
Clinically presents as tense bullae located especially the trunk and perineum. It mainly affects young children.
We present the case of a female child of 35 months who came to our consultation presenting a one-month history of a pruriginous vesicobullous eruption. On physical examination she presented tense bullae with translucent content on erythematous and normal-appearing skin, symmetrically distributed over her palms and soles and circinated erythematous patchs with peripheral blistering, and crusting on the perioral area, neck and anterior thorax. In 24 hours those lesions evolved, involving the thorax and the back and presenting as annular or circinate lesions with central clearing and blistering at the edges, resembling a "string of pearls".
A punch biopsy was taken. It revealed findings compatible with IgA linear dermatosis.
She began treatment with prednisolone (1mg/kg/day) and dapsone, responding only partially, reason why intravenous immunoglobulin (IVIG) were added.
Systemic corticosteroids were gradually reduced until suspension. IVIG were suspended and flucloxacillin was added. She is now medicated with dapsone (2mg/kg/day) and flucloxacillin with only a parcial response.
With this report we pretend to present a case of a rare dermatosis with a chronic evolution, characterized by remissions and relapses and a difficult therapeutic management in most of the cases.
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PP093 - LICHEN PLANUS: OUR EXPERIENCE OF 106 CHILDREN

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Background: There is scarcity of studies on LP in children.
Objective: To delineate epidemiology, clinical features, precipitating factors and treatment pertaining to childhood LP.
Patients and methods: In this retrospective study, records of children with LP registered at the pediatric dermatology clinic of our center, between January 2001 and April 2008 were reviewed. The diagnosis was made on clinical grounds. Information regarding age, sex, duration of disease, initial site of onset, areas of involvement, morphology of lesion nail involvement, mucosae or scalp involvement , associated diseases, family history, history of viral hepatitis, history of hepatitis B vaccination or any other apparent precipitating factor(s), treatment offered and response to treatment were obtained.
Results: Mean age of the patients at the time of consultation

was 8.75 years, while mean age at the onset of the disease was 7.55 years. Lower limbs including feet were the commonest site of onset in 55%patients. History of hepatitis B vaccination was there in 20.8% patients. Classical papular lesions was the commonest morphology observed in 43.4% patients followed by eruptive, linear, isolated nail involvement, LP hypertrophicus(LPH), actinic LP, lichen planus pigmentosus and bullous LP. Oral mucosal involvement was observed in 16.9% patients while nail involvement was observed in 18.8% patients.
Oral prednisolone (1mg/ kg) was given to those who had extensive/ eruptive lesions. Topical corticosteroids remained the treatment of choice in most patients with localized lesions.
Data of at least one follow- up visit at least 4 weeks after starting treatment was available in 22 patients. The mean follow- up duration was 18.1 weeks. All patients with follow- up data had either improvement in their skin condition or complete clearance with treatment. Post- clearance follow- up data was not available in any patient.
Conclusions: In the largest ever study on childhood LP, we found that it occurs apparently more commonly in males, nail involvement may not be that uncommon as in commonly thought, and a significant percentage of patients have a history of previous hepatitis B vaccination.

PP094 - ETANERCEPT IN PEDIATRIC PSORIASIS: DEFINITION A NEW HETEROGENEITY IN COST EFFECTIVENESS CONCEPT, IN DEVELOPING COUNTRIES: EVIDENCE BASED PEDIATRIC DERMATOLOGY APPROACH

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Objective: Biologic agents have been designed with the help of immunological studies to target particular areas of the immune system which are thought to play a role in the pathogenesis of Disease. Etanercept is a soluble, anti-tumor necrosis factor alpha
(TNF-α) agent is used for the treatment of adult and recently pediatric psoriasis. We evaluate the efficacy of etanercept in various subtype of pediatric psoriasis.
Patients and methods: Recalcitrant generalized psoriatic Patients 4 to 17 years old received 0.4 mg of etanercept per kilogram of body weight subcutaneously, twice a week for up to three months in the initial, Open-label part of a trial. Cost effectiveness was evaluated by recommendation forms that completed by patients and their parents separately in the cases eldaer 7years.
Results: All the 5 patients with generalized recalcitrant pustular psoriasis experienced marked and more important sustained improvement but our finding about generalized recalcitrant plaque was disappointing.
Improvement in one of them was evaluated less than 50% and in the other patients no improvement were recorded. Cost effectiveness calculation were lead to various results in two patients between patients and parents. Mean of follow up duration were .5.3 ± 1.7months
Conclusion: Based on my findings generalized pustular psoriasis is suitable type for the treatment by etanercept; although regarding to drug price all the patients were treated only up to 3 months. Cost effectiveness heterogeneity in this study is due to incomplete insurance coverage that lead to more distress in parents and showed this variable in Pediatric Dermatology should be considered separately at least in developing countries.

PP097 - PEDIATRIC PUSTULAR SWEET SYNDROME

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Introduction: Sweet syndrome or acute febrile neutrophilic dermatosis, is a rare entity reported in children. It is characterized by the presence of erythematous and edematous plaques, accompanied by high fever, neutrophilia, a dense neutrophilic infiltrate in dermis and an excellent response to systemic corticosteroids.
Case report: A four-year healthy spanish boy was admitted to hospital for the presence of an erythematous and edematous plaque on the left upper eyelid which began four days before, related to an insect bite by his parents. It was accompanied by the later appearance of edematous plaques and pustules on the posterior neck, eyelids and face, as well as high fever. He began treatment with oral and topical antibiotics, with no response. Inmediately, new erythematous and edematous plaques start to appear on the trunk and extremities. We took cutaneous cultures, performed Tzanck stain, pharyngal smear and hemocultures which were all negative and we made a skin biopsy. The hemogram showed discrete leukocytosis with neutrophilia and elevated C-reactive protein. After four days of clinical worsening and with Sweet syndrome suspect, we add systemic corticosteroids to his treatment with a dramatically good response .The biopsy revealed a mixed panniculitis, specially composed by neutrophils. All the tests carried out to rule out malignancies were negative. Blood tests only demonstrated IgG for herpes virus type 6 and elevated ASLO.
Conclusion: We report a case of Sweet Syndrome with the presence of facial vesicles and pustules, which is an unusual type of pediatric presentation.

PP098 - PROFILE OF PITYRIASIS LICHENOIDES IN CHILDREN- IS IT SELF LIMITING?

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Background- Pityriasis lichenoides (PL) is a T-cell mediated eruption of unknown etiology. There are no established guidelines for its treatment(2).
Objectives- Studies on childhood PL are few and its course and treatment are obscure.
Materials and Methods- A prospective,open, clinico-investigative study of 10 children (<18 yrs) with PL.
Observations- There were 7 boys (70%) and 3 girls(30%) in the age group of 2.5 – 18 years, mean age (7.2 yrs). Duration varied from 2months - 3 years. Associated diseases were - atopic dermatitis in (1), hand eczema in (1) and HIV infection in 1child. PLC (Pityriasis lichenoides chronica) was the commonest presentation in 8 children (80%). Presence of papulo-vesicular lesions, crusted plaques and scars in addition to PLC were noted in 2(20%), and were diagnosed as

PLEVA with PLC. Trunk and extremities were involved in all. Eosinophilia in (3), raised Ig E in (2) and deranged liver function tests were noted in (1) child . Emollients, topical steroids and tacrolimus ,erythromycin, PUVAsol and NB-UVB were used for treatment with unsatisfactory response in 6 (60%) children. Relapses were common.
Conclusions- PLC is more common children. The two can occurtogether. Role of eosinophilia and atopy requires further studies. Treatment is frustrating. Self limiting course is doubtful.
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PP099 - A COMPARISON BETWEEN EUROPEAN AND ASIAN CHILDREN WITH PSORIASIS

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Background. A comparison between European and Asian children with psoriasis has never been performed.
Objectives. To study the epidemiological characteristics of childhood psoriasis in the Netherlands and Singapore and to compare between these patients.
Methods. Data were collected from juvenile psoriasis patients (<18 years old) in the two countries. Dutch data (NL group) were collected prospectively at the dermatology outpatient clinic at the RUNMC. Retrospective data were obtained from the casenotes at NSC for the Singaporean cohort (SG group).
Results. The NL group consisted of 39 patients and the SG group had 162 patients. Female:male ratios were: 1.3 : 1 (NL group) and 1.4 : 1 (SG group). The mean age of onset was 7.1 years (0.5-14.1) in the NL group and 10.8 years (0.5-16.9) in the SG group. A positive family history was obtained in 71.8% of NL group and 13.6% of the SG group. The majority of Dutch children (84.6%) experienced itch whereas only 14.2% of the Singapore children did. In both groups, plaque psoriasis was the most common type (79.5% NL group; 55.6% SG group). In Dutch and Singaporean patients guttate psoriasis was rare (2.5% NL group;4.7% SG group). Pustular psoriasis was uncommon in both groups. The scalp was the most common site affected in both groups (92.3% NL group;67.9% SG group) followed by the limbs (87.2% NL group;46.9% SG group). There was a similar proportion of children with nail changes in both groups (23.1% NL group; 35.8% SG group).
Conclusion The female:male ratio, sites affected and types of psoriasis were similar for both groups. A striking difference was found in the incidence of positive family history and reporting of itch. This study demonstrates interesting differences between European and Asian juvenile psoriasis patients.

PP100 - EFFICACY AND SAFETY OF TREATMENTS IN CHILDHOOD PSORIASIS: A SYSTEMATIC LITERATURE REVIEW

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Background. Evidence-based recommendations for therapeutic decision-making in juvenile psoriasis are lacking.
Objectives. To systematically review all available literature concerning treatment efficacy and safety in childhood psoriasis and to propose a recommendation for topical and systemic treatment of childhood psoriasis.
Methods. Databases searched were Pubmed, EMBASE, and the Cochrane Controlled Clinical Trial Register. All studies reporting on efficacy and safety of all treatment options in childhood psoriasis were obtained and a level of evidence was determined.
Results. Literature search revealed 2649 studies, of which 64 studies met the inclusion criteria. The majority of topical and systemic therapies given in juvenile psoriasis are efficacious. Short-term side effects were usually mild; long-term side effects were not described.
Conclusions. The following treatment algorithm is based on the results of the review. The treatment of choice in mild or moderate juvenile psoriasis is calcipotriene, if necessary, combined with mild to moderate topical corticosteroids. In case of treatment-resistant flexural and/or facial psoriasis, tacrolimus 0,1% can be added to the treatment regimen. If this treatment regimen is not effective, or if psoriasis is moderate to severe, treatment with dithranol is recommended. Only in case of lack of efficacy of these modalities, treatment with NB-UVB can be considered in adolescents, but only for a short duration. Of the systemic treatments, MTX is regarded as the therapy of choice. Retinoids should be considered in cases of pustular and erythrodermic psoriasis. Treatment with cyclosporin should only be deliberated in exceptional cases. Etanercept is a very promising new treatment modality, which should be considered as a third-line drug.

PP101- TREATMENT WITH ACITRETIN IN CHILDREN WITH GENERALIZED PSORIASIS: EXPERIENCE IN THREE PATIENTS

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Background. About one-third of psoriasis vulgaris cases seen in the pediatric age group. The need for safe, effective therapy is a special consideration in pediatric population.
Objectives. To document our preliminary experience with acitretin in the pediatric patient with generalized psoriasis
Methods. Three pediatric patients with extensive generalized psoriasis were treated with acitretin 0,5 mg/kg/day for 6 months. Laboratory tests include liver function, serum lipids were performed before treatment and one–month interval. Also baseline and six-months bone studies considered. Neurologic and ophthalmologic examinations were performed before and after treatment.
Results. Three patients, two girls and one boys, 9 to 13 years of age, with PASI scores of 15,7 and severe area affected by psoriasis 40%. After three months of treatment, they all had complete clearance of erythema and scaling. At the end of the treatment, they all achieved 6 months treatment with

90 % improvement of the PASI score. Scalp area of the patients showed minimal resistance to the Acitretin therapy. Advers events are limited and transient; cheilitis, pruritus and dryness were observed. There were no detected any advers effects in liver function and serum lipid levels during the therapy.
After six months of the acitretin therapy remission of the diasease were continued.
Conclusion. This study is limited by number and age range, but our observations suggets that acitretin is effective and well tolerated drug for the treatment of severe forms of generalized childhood psoriasis before other systemic drugs such as methotraxate and biologic agents.
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PP102 - CHILDHOOD PSORIASIS: REVIEW OF 56 CASES

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Objectives: To register the epidemiologic and clinic characteristic of infantile psoriasis, to analyze the different therapeutic modalities used and compare our data with other published series.
Methods: retrospective study of the patients with psoriasis admitted in the consultation of Pediatric Dermatology of the Hospital de Braga (Braga, Portugal), between the years of 2001 and 2008. The following parameters had been evaluated: sex, age in the first consultation and first manifestation, familiar history of psoriasis, history of exuberant diapers dermatitis, precipitation factors, clinical form, localization, pruritus and therapeutic measures.
Results and discussion: During these 8 years 56 children with psoriasis had been admitted in the consultation of Pediatric Dermatology. There were 25 (44.6%) boys and 31 (55.4%) girls, with a male to female relation of 1:1.24. The mean age of the first consultation was of 9,7 years and of the first manifestation was 7,3 years. Girls showed an onset of psoriasis more precocious with a peak between 4-6 years (32.2% of the children of the feminine sex). Classical plaque psoriasis was the most frequent clinical presentation (71.4%), followed by guttata form (23.1%). A positive familiar history was present in 41% of the cases and 19.6% of the children had antecedents of exuberant diapers dermatitis. In the majority of the patients (73.2%), the control of psoriasis was achieved with topical measures, 13 children (23.2%) were treated with phototherapy and 15 patients (26,8%) with systemic treatments (21,45 with cyclosporine 21,4%, 7,1% with acitretin,1,8% with methotrexate, 7,1% with etanercept and 3,6% with infliximab).

PP103 - PSORIASIS: A COMMON BUT SOMETIMES DIFFICULT DIAGNOSIS

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Case report: A 16-month-old male toddler presented a 10-month evolution intensely itchy eruption comprised of scaly papules and coin-shaped plaques, some of them covered with honey-colored crust, symmetrically distributed over the trunk and limbs. The family history was irrelevant, except for the presence of dogs at home.
The diagnosis of bacterial overinfection of atopic dermatitis was proposed. He was treated with topical steroid, oral antibiotic and antihistamine without improvement. Oral steroid was added with no significant change. Other diagnoses were proposed, including dermatitis herpetiformis and fungal infection. KOH test was suspicious and blood studies, including CBC, biochemistry, immunoglobulin levels, antigliadin and antitransglutaminase antibodies, specific IgE to food and aeroallergens, and autoimmune panel were normal or negative. Gluten-free diet and oral griseofulvin were tried with no clinical alteration.
Skin biopsy showed epidermal parakeratotic hyperkeratosis, irregular acanthosis, cellular debris forming crust, spongiosis predominantly suprapapillary and neutrophil exocytosis, often in intraepidermal clusters (impetiginized eczema according to the pathologist). The eruption progressively extended to the face and scalp and lesions acquired a more whitish-silvery scale as bacterial superinfection resolved. The clinical appearance and evolution of the lesions, the poor therapeutic response and some histological clues suggested the diagnosis of psoriasis. Treatment was tried with topical betamethasone-calcipotriol ointment with progressive improvement.
Comments: Psoriasis is a common condition. In childhood, the clinical pattern of psoriasis often takes on a peculiar form and may differ from adult psoriasis in its presentation and treatment. Determining the correct diagnosis is essential for effective treatment, as showed in our patient.

PP104 - ANNULAR LICHENOID DERMATITIS OF YOUTH: TWO FURTHER CASES

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Annular lichenoid dermatitis of youth (ALDY) is a rare entity described first by Annesi et al in 2003. It is a lichenoid dermatitis of which etiology is unknown, although one case after hepatitis B vaccination has been reported. Here we report two further cases of ALDY. A 12 year-old boy presented with asymptomatic hypopigmented lesions on the abdomen and legs of 1 year duration.The lesion on the abdomen had an erythematous annular rim. Annular lichen planus, morphea, hypopigmented mycosis fungoides and figurate erythema were the presumptive diagnoses though the lesions were not characteristic for any. Histopathologic examination revealed hydropic degeneration and apoptotic cells at the basal layer of the epidermis, together with a bandlike primarily lymphocytic infiltrate in the upper dermis with some melanophages. With the clinicopathologic correlation, diagnosis of ALDY was established. Lesion on the abdomen resolved

with the use topical corticosteroid for 2 weeks leaving hypopigmentation. The second case was a 7 year-old girl with a single asymptomatic hypopigmented lesion with an erythematous rim located on the abdomen for 1 year duration. Histopathologic examination lead to the diagnosis of ALDY. With narrowband UVB and topical corticosteroid lesion resolved leaving hypopigmentation. The cases are presented to emphasize the self-limited nature of this benign dermatitis which is probably insufficiently known.

PP105- PITYRIASIS ROSEA IN TWO SISTERS

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Background: Pityriasis rosea is an acute, inflammatory dermatosis frequently affecting adolescents and young adults. It's current name originated from Gibert in 1860 year. It is common, self-limiting, papulosquamous disorder affecting the trunk and limbs.
Objectives: Occuring most often during the spring and autumn, pityriasis rosea is believed, although not proven, to be of viral origin. Although generally asymptomatic, it may vary from mildly to severely pruritic. Familial disease pattern is not common; when encountered, it raises the suspicion of infectious etiology.
*Method*s: We here present two patients, 17 and 12 years old sisters. The first child (older one) was presented with the eruption on the neck, upper parts of the arms and lateral trunk aspects. Nine days later, similar disease occurred in younger sister. The lesions were tipical, numerous, oval plaques (the biggest one was 15mm in diameter) with central, wrinkled, pale area and redish peripheral zone with fine scaling. In both patients there were no significant subjective complaints and primary medallion didn't appear. Younger sister had also a moderate lymphadenopathy in axillar regions.
Results: Both patients was given oral therapy with acyclovir and topical therapy with aclomethasone, so as an advice to avoid extensive bathing and irritation. After one month, there were no symptoms left.
Conclusion: The patients had also one more sister (16 years old) and a brother (14 years of age), but they didn't have any symptoms of the disease. So, the cause of the disease remained unclear.

PP106 - BIOLOGIC THERAPY IN SEVERE CHILDHOOD PSORIASIS

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Psoriasis is a common, chronic, disfiguring, inflammatory and proliferative condition of the skin. It is not rare disease in the pediatric population, which is immunologically mediated, caused by activation of T-lymphocytes that elaborate a Th1 type of immune response. As the complex molecular mechanisms underlying the pathogenesis of psoriasis become increasingly clear, targeted therapies aimed at specific components of the inflammatory cascade, such as tumor necrosis factor, are gaining popularity.
According to these new findings, in moderate-to-severe cases, treatment is challenging. Recently, there have been advances in the use of biologic therapies, specifically tumor necrosis factor (TNF)-alpha blockers, for pediatric psoriasis. Though the cost of the biologics is a limiting factor, their unique action has definitely given a new hope for the management of psoriasis.
In Hungary for childhood psoriasis the biologic therapy is available since 2009. Only etanercept is formally approved for the treatment of psoriasis under 18 years by the Hungarian Association of Dermatologists. The biologic therapy should be kept reserved for severe psoriasis only. According to the guidelines, the patients are required to meet all these conditions: severe disease defined as PASI score of 10 or more (or a BSA of 10% or greater where PASI is not applicable) and a DLQI (dermatology life quality index) >10. The course of the disease should be severe for at least 6 months, resistant to one of the systemic treatment (acitretine, UVB phototherapy, cyclosporine) and should fall in one of the well-defined clinical categories.
The first and single center for biologic therapy in childhood psoriasis takes place in our Department.
The authors would like to report about the promising experience.

PP107 - ACRODERMATITIS ENTEROPATHICA: A NOVEL SLC39A4 GENE MUTATION FOUND IN A PATIENT WITH AN ATYPICAL PRESENTATION

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Introduction: Acrodermatitis enteropathica (AE) is a rare autosomal recessive disorder caused by a defective absorption of zinc. Usually the symptoms emerge in infants who are not breastfed. After identification of the AE SLC39A4 gene in 2002, a few mutations of this gene in AE have been described in literature.
Case report: A 3-month-boy, full-term and breast-fed, presented with erythematous-erosive lesions localized in perioral, perianal, and acral areas. There was no diarrhea or alopecia. The parents were consanguineous but importantly there was no family history of dermatologic disorders.
His serum zinc level was low (0.34 mg/L; normal 0.70-1.20), as well as the serum alkanine phosphatase. The mother's serum zinc was normal (0.94 mg/L), but the breast milk zinc was decreased (0.36 mg/L; normal at 12 weeks: 1.35 mg/L).
With the hypothesis of AE, replacement with oral zinc was initiated and a rapid improvement of the cutaneous lesions was appreciated.
Mutation analysis performed in the child revealed a homozygous insertion in exon 3 in SLC39A4 gene.
At present, the child is 3 years and is still under zinc replacement, with no cutaneous lesions.
Discussion: Prompt recognition of this disorder is essential in order to initiate zinc supplementation and prevent the long term consequences of zinc deficiency. In this case the authors emphasize the onset of AE in a breast-fed child, and the report of a mutation never described before. This is an additional contribute for the knowledge of the genotype-phenotype correlation and the molecular basis of AE.

PP108 - PSORIASIS IN PEDIATRIC POPULATION – EPIDEMIOLOGIC, CLINIC AND TREATMENT PARTICULARITIES IN A FOUR YEARS SURVEY

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Psoriasis most often begins after puberty, but in 25-45% of patient the disease begins before the age of 16 years, in 10% before the age of 10 years and in 2% before the age of 2.
In respect of the pediatric population, the disease has an important impact on the child and family QoL and in addition to dermatological care, attention must be given to the emotional problems that may be created in the child and family.
125 children and teenagers were enrolled in our survey over a period of four years (January 2003 – December 2006), representing 9.72% of the total number of 1286 children and teenagers referred to our Pediatric Department. The sex ratio was male to female 1.4:1, median age being of 9.2 years.
With regard to the clinical form of psoriasis we found: plaque psoriasis (39.2%), guttate psoriasis (21.6%), palmoplantar (8%), scalp (4%), and erythrodermic (4%), nail psoriasis (2.4%)
Regarding the most often involved areas we noticed extremities (42.4%), scalp (39.2%), body (35.2%).
In 60% of the cases the treatment was topically applied, in 38.4% UVR were associated and 1.2% needed systemic therapy.
Our results were compared to other statistic data and we noticed useful new information emerged from this effort in respect of the diagnostic and treatment of the disease in children and teenagers: for the clinical diagnostic attention should be addressed to the scalp and nail lesions and to small guttate lesions, too; for the therapeutic approach it is useful to involve a psychologist in the team to improve the psychological status of the patient and family members.

PP109 - PITYRIASIS RUBRA PILARIS – REPORT OF A CASE TREATED WITH ISOTRETINOIN

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Pityriasis rubra pilaris is a chronic inflammatory dermatosis of unknown etiology, with a large clinical variability.
We report the case of an 11 year-old male child who presented with a week history of ill-defined erythema and fine scaling of the face, erythematous follicular papules, some of which joined in well-defined plaques, located to his elbows and sacred region and orange keratoderma of the palms and plants.
A punch biopsy was made and their findings supported the clinical diagnosis of pityriasis rubra pilaris. He started treatment with oral isotretinoin (0,75mg/kg/dia) and 2 months after all lesions had resolved.
Pityriasis rubra pilaris is a rare dermatosis, mainly in children. A universal treatment doesn´t exist, although systemic retinoids are the most accepted one.
With this report the authors intend to present a rare condition, warning of its clinical variability and therapeutic options.
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PP111 - TO DIE OR NOT TO DYE!

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Henna tattoos became fashionable in the West in the late 1990's as a form of temporary body decoration and have since become extremely popular, especially with children on holidays. Natural henna is reddish brown in colour and its application is rarely associated with problems. Holiday resort henna is frequently black due to the addition of phenylenediamine (PPD) which makes the tattoo easier and quicker to apply.
PPD is a chemical substance that is also widely used in permanent hair dye and has been associated with both delayed-type and immediate-type hypersensitivity reactions, including fatal anaphylaxis. There are also reports of cross-reactivity to PPD, with certain sunscreens, some local anaesthetics and several medications.
We describe a case of a 12 year old boy who presented to hospital with a life threatening allergic reaction 4 days after dyeing his hair at home. Of note 18 months before this he experienced a reaction to a 'black' henna tattoo resulting in scar formation at the site of application. He also gave a history of an allergic reaction to sunscreen.
With the increasing popularity of henna tattoos and the increasing use of hair dyes in the paediatric and adolescent population there is a need to increase awareness amongst professionals and educate the public with regard to this potentially fatal complication of a seemingly safe practice.

PP113 - AN ECZEMA EDUCATION PROGRAM FOR GENERAL PRACTITIONERS, DOES IT INCREASE LOCAL EXPERTISE AND IMPROVE TIMELY ACCESS TO TERTIARY SERVICES?

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Background: Eczema is a chronic and debilitating disease with a risk of many associated negative outcomes. The approximate waiting time for non urgent patient to be seen in The Royal Children's Hospital (RCH), eczema clinic (EC), Australia is greater than 8 months. Most referrals are from General Practitioners (GP) and skin disorders are one of the most commonly seen conditions in General Practice[1].
Objectives: To increase the knowledge and confidence of GP in the effective treatment of mild to moderate eczema and in turn improve timely access for patients to tertiary services.
Method: A program was developed and provided to 46 GP. It consisted of three parts; firstly a self directed pre activity exercise, secondly an evening interactive lecture and thirdly attendance at a RCH EC. The programs effectiveness was assessed by; referral patterns from the participating postcodes,

the waiting time for a new non urgent patient to be seen in the EC, an evaluation of the rate of patients failing to attend dermatology clinic appointments, participant knowledge, confidence and satisfaction and evaluation of the program.
Results: Participants were extremely satisfied with the program. There was an increase in their knowledge and confidence in treating eczema. There was no significant change in the waiting time or patient failure to attend rate in the dermatology clinics or the number of new referrals from GP to the dermatology clinic.
Conclusions: The program was very successful in improving GP knowledge and confidence in managing mild to moderate eczema. It is imperative to include Divisions of General Practice in the planning, advertising and organization and also for the program to be accredited with Continuing Professional Development (CPD) points.
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PP115 - COMBINATION OF MALASSEZIA TO HOUSE DUST MITE AS AN IMMUNOMODULATOR: DEFINITION A NEW MODEL BASED ON TRANSLATIONAL RESEARCH

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Background: Atopic dermatitis (which is synonymous with atopic eczema) is an itchy, chronic or chronically relapsing, inflammatory skin condition. Dermatit atopic management is very difficult and there is various options.It seems that immunomadulators are an rational option in the Atopic dermatit management.we intend to consider the combination of Malassezia to house dust mite as an immunomodulator based on translational research principles.
Materials & methods: In AD Naive precursor Th(T helper)0 cells are induced to differentiate into Th2 cells, characterized by the production of interleukins
(IL) -4, -5 and -13. Th2 cells 'help' or control the type of immunoglobulin (Ig) that B lymphocytes make, inducing synthesis of IgE.I use of Th cells as the first axis.
There is accumulating evidence that DCs(dendritic cells) may also subdivide into DC1, preferentially inducing Thl cells,and DC2, inducing Th2 cells.
The maturation of DC will lead to surface marker expression and cytokine production, as revealed by upregulation of the costimulatory molecule CD86 and production of CXCL8. Actually, without such upregulation, DCs may
instead exhibit tolerogenic properties, preventing the generation of effector T cells.I use them as the second axis
Results: Based on my model critical point is that desensitization with allergen ,especially house dust mite,may lead to DC 2 pathway.if we use of malasezzia antigen as a ligand to allergen, DC induce naïve Th to Th 1 instead of Th2. So allergen will be recognized as an ordinary and foreign antigen. Meanwhile we will be sure that immune system could able to overcome Malassetia and no adverse complication encountered.cost benefit ratio calculation showed this pathway is more rational in compare to other alternatives such as Mycobacterium vaccae. Suspensions and probiotic suspensions added to milk formulas in infants.
Conclusion: This model help us to draw new viewpoints in DA management based on translational research principles and improve our evidences.

PP116 - PRO INVESTIGATION: PREVALENCE AND ORIGINS OF HYPERSENSITIVE SKIN AMONG MORE THAN 8 000 CHILDREN

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Aims: Children with Hypersensitive skin (HS) represent a reality but very few data are available. An international epidemiological study has been conducted in France, Spain, Italy and Taiwan in 2009.
Methods: Paediatricians filled firstly a register of all children <6 years old, and secondly a questionnaire about HS children, their family, triggering factors, clinical signs and management. Exclusion criteria were Atopic Dermatitis and allergic contact dermatitis.
Results: Among 8112 children included in registers, 2482 (30.6%) were declared with HS (50.4% boys). 13.1% were < 3 months old and 34.9% >24. HS begins often early in life: 21.3% at birth. Familial HS is present in 56.4% of parents (71.1% mothers) and in 54.7% of brothers or sisters. Triggering factors are various: environmental (heat 44.1%, cold 35.3% , temperature variations 33.1%), mechanic (sweat and physical effort 48.6% , scratching 40.2%, contact with synthetic material 31.9% and wool 26.3%), external (products in contact with skin such as cosmetics or washing powder 68.9%, hard water and swimming-pool 65.8%) and/or psychological (intense emotion 77.2%).
Visible cutaneous signs are present on the face (62.5%) and body (65.7%), mostly erythema (73.3%) and dryness (61.8%). Symptom is mostly pruritus (36.6%). Flares last long: 54.8% several days and 20.9% a few hours. 65.4% of parents have applied cosmetics products to improve this HS.
Conclusion: HS affects 30% of children in Europe and Asia. It begins very early in life and has a familial trait in about half of children. This study, which demonstrates specificities among different countries, provides first clues for management.

PP117 - HYPERSENSITIVE SKIN IN CHILDREN : SPECIFIC SKIN CARE PRODUCTS

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Background: Sensitive skin is becoming a prominent problem among adults. It has never been studied in the paediatric population and there are no specific skin care products for hypersensitive skin (HS) in children. Four products have been conditioned under protective atmosphere, with no perfume, no preservative. They contain patented Avocado Su-

gars (AV) which help to preserve the skin by enhancing the structural organization of the epidermis (collagen VII; beta-4 integrin; involucrin; desmoglein 1) as well as the antimicrobial barrier (Human Beta-Defensins).
Methods: Four European multicenter open-label tests involved 78 dermatologists and pediatricians in France, Spain and Italy. During April and May 2007, they included children (maximum age 7) with HS, assessed by the accompanying person (mother or father).
After clinical examination, tested products (Face cream, Body milk, No-rinse cleanser and Cleansing foam) had to be use under normal conditions for 21 days and a questionnaire was sent at the end of the 4 studies, concerning efficacy, tolerance and cosmetic qualities.
Results: 531 children, mean age 2.8, were included. 26.7% had normal appearing skin, 27% Atopic Dermatitis, 47% Irritant Contact Dermatitis, 38% Dry skin, and 16% Perioral Dermatitis. 49% had a sensitive skin familial background and 30% and atopic one.
Global results were respectively for efficacy, tolerance, cosmetic qualities:
Face Cream (118): 88%/100%/98%; Body Milk (129): 83%/95%/93%; No-rinse Cleanser (135): 83%/94%/91%; Cleansing Foam (149): 93%/95%/88%.
Conclusion: The concept of sensitive skin is applicable to children and specific skin care products are needed. As for adults, they must protect the skin and be perfectly tolerated.

PP118 - NATURAL PPAR-ALPHA AGONIST AND ATOPIC DERMATITIS: FROM RESEARCH TO CLINICAL EFFICACY

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Introduction: Natural Peroxysome-Proliferative-Activated Receptors ligands are able to restore skin barrier function, demonstrate anti-inflammatory properties and have a potential role in Atopic Dermatitis (AD) management. An emollient containing a patented Sunflower Oleosdisitillate (SO) has been formulated for AD skin care.
Protocols: *It has been demonstrated that SO:
- Activates PPARs-alpha, induces epidermal key lipids synthesis and reduces cutaneous inflammation.
*Study A: Randomized study on the Steroid-sparing effect and Quality of life (QoL) impact of 2% SO. 86 children (4-48 months) with AD have been attributed to 5 different treatment groups during 21 days: desonide 0.05% 2X/D to 1D/2, with or without 2% SO cream.
*Study B: Randomized study comparing 2% SO cream 2X/D to hydrocortisone butyro-propionate (1 mg/g) 2X/D for 21 days among 40 children (mean age 2.3 years), with moderate AD (mean SCORAD 37).
Results: *Study A: Similar and significant improvement of SCORAD ($p<0.01$) in all groups: 63% in the TS 2X/D group and 75% in the TS 1D/2 + 2% SO cream group. QoL improvement was at best in the 2% SO cream groups.
*Study B: In the two groups, SCORAD decrease was significant at D7 and D21 (-70% and –75%), with no statistical difference of SCORAD at D21 (11 vs 9.4). QoL questionnaires were similarly improved in both groups (65 to 75%).
Conclusion: For the first time, a cream containing Sunflower Oleodistillate has demonstrated comparable therapeutic properties to a Topical Steroid in a randomized comparative study during 3 weeks. These properties are probably linked to previously demonstrated agonist PPAR-alpha properties.

PP119 - EFFICACY AND SAFETY OF AN EMOLLIENT CONTAINING PENTYL RHAMNOSIDE IN ATOPIC DERMATITIS: AN OBSERVATIONAL SURVEY ON 649 ATOPIC CHILDREN

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Objective: The aim of this observational study was to observe the improvement of atopic dermatitis after using an emollient balm or cream containing pentyl rhamnoside during 3 months.
Methods: This observational study was conducted in six countries.
The patients applied an emollient balm or cream twice a day to the areas of atopic dermatitis, excluding areas of inflammatory flare-up for 90 days.
The primary efficacy endpoint was the severity of atopic dermatitis assessed by the SCORAD at D90.
The secondary efficacy endpoints included the number of flare ups, the severity of subjective and objective clinical signs and the quality of life. The overall satisfaction and tolerance were also evaluated.
Results: This observational study was conducted upon 649 atopic children.
After 3 months of application, a significant improvement of SCORAD (-65%, $p<0.0001$) was noted.
The number of inflammatory flare-ups was significantly decreased (-47%, $p<0.0001$).
A significant improvement of all clinical signs was also observed (ranging from -65% to -68% after 3 months, $p<0.0001$ versus D0).
An improvement of quality of life of 67% ($p<0.0001$) was also reported.
The global efficacy and the tolerance of the treatment were satisfying or very satisfying for more than 90% of investigators and patients.

PP120 - NURSE'S ROLE IN PREVENTION OF DIAPER DERMATITIS

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Introduction: Diaper dermatitis is a highly prevalent condition that causes discomfort and stress for patients and frustration for nurse staff. The skin is an interface for primary care in any patient-caregiver interaction.
Diaper Dermatitis is the most common dermatological problem of infancy, occurring in 25 – 65 % of children and is

caused by the combined effect of irritants such as diaper, urine and feces.
Discussion: Studies have shown the best thing to do is prevention. Diaper technology has evolved to substantially lessen the severity of diaper dermatitis, but additional improvements are needed. Premature infants and incontinent adults are particularly at risk for developing diaper dermatitis and its potential consequences. Contributing factors include over hydration, irritants, friction, increased skin pH, diet, gestational age, antibiotic use, diarrhea and medical condition. Treatments aim to reduce hydration, provide a semi permeable 'layer' to facilitate skin barrier repair, shield the skin from irritants, deactivate specific fecal components and maintain skin surface contact. However prevention of diaper dermatitis can be summarized with the acronym ABCDE (air, barrier, cleansing, diaper, and education). Provide education regarding diaper dermatitis to parents or caregivers is important. If treated using the ABCDE acronym, the prognosis is excellent too.
Conclusion: As primary prevention is important, it is necessary to pay attention to Education.Providing education to the parents and/or caregivers is important in the treatment and further prevention of diaper dermatitis.Nurses have a key role in effective care and educations,the nurse's accurate assessment,promote intervention and evaluations are necessary for better outcomes.

PP121 - IL-31 EXPRESSION BY INFLAMMATORY CELLS IS PREFERENTIALLY ELEVATED IN ATOPIC DERMATITIS

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Background: Interleukin-31 (IL-31) is a recently discovered cytokine expressed in many human tissues, and predominantly by activated CD4+ T cells. IL-31 signals through a heterodimeric receptor consisting of IL-31 receptor alpha (IL-31RA) and oncostatin M receptor beta (OSMR). Earlier studies showed involvement of IL-31 and its receptor components IL-31RA and OSMR in atopic dermatitis, pruritus and Th2-weighted inflammation on the mRNA level.
Objective: Investigate IL-31 protein expression in atopic dermatitis, Th2-weighted and pruritic skin diseases to explore the extent of IL-31 involvement in such conditions.
Methods: Immunohistochemical staining for IL-31, IL-31RA and OSMR of the inflammatory infiltrate in formalin-fixed paraffin-embedded biopsy specimens of Th1-, Th2-weighted, pruritic and non-pruritic skin diseases.
Results: IL-31 expression was increased in the inflammatory infiltrates from skin biopsies taken from subjects with atopic dermatitis, when compared to controls (p=0.05 and below). IL-31, IL-31RA and OSMR protein immunoreactivity was not increased in biopsies from subjects with other Th2-weighted and pruritic skin diseases.
Conclusion: Our results confirm, at the protein level, the relationship between IL-31 expression and atopic dermatitis that was previously identified at the mRNA level. Our results do not support a general relationship between expression of IL-31/IL-31R and pruritic or Th2-mediated diseases.

PP122 - CLINICAL EFFICACY AND SKIN TOLERABILITY OF A NEW SKINCARE PRODUCT (EUCERIN(R) SKIN CALMING LOTION 12% OMEGA) IN CHILDREN WITH ATOPIC DERMATITIS

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Atopic dermatitis is characterized by a disturbed epidermal barrier (deficiency of ceramides and omega-6 fatty acid) and a high colonization density of the skin with Staphylococcus aureus that correlates directly with the severity of the skin disease. In this study it was investigated whether consistent skin care with an emollient lotion with a high content of omega-6 fatty acids leads to an improvement in the atopic skin condition.
The two-week treatment with 12% Omega lotion led to a clear improvement in the skin condition in over 80% of the children. Especially the parameters dryness, scaling, erythema and itching were improved. The SCORAD index decreased by more than 50% overall relative to the baseline finding.
The improvement in skin condition correlated with a clear reduction of Staphylococcus aureus colonization density. The skin tolerability of the product was assessed as "very good" to "good" by over 80% of the patients and the product properties were consistently assessed positively. The product is therefore suitable not only for adjuvant care but also for preventive care in symptom-free intervals.

PP123 - PHOTOSENSITIVITY TESTING IN CHILDREN

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Background: Phototesting is an important diagnostic tool to objectify light-related symptoms. Data on phototesting procedures in children are scarce.
Objective: The aim of this study was to evaluate phototest results in photosensitivity disorders in children.
Methods: The phototest procedures are described. All children phototested in our department between 1995 and 2007 were included in this retrospective study. Children diagnosed with polymorphic light eruption were selected for follow-up.
Results: A total of 92 children (39 boys and 53 girls, age range 4-16 years old) were successfully phototested. A photosensitivity disorder was confirmed in 56 children (61%, 24 boys and 32 girls). Polymorphic light eruption (PLE) was diagnosed in 39%, photosensitivity associated with atopic dermatitis in 23%, and erythropoietic protoporphyria in 23%. Other diagnoses were less common. Ten children with PLE were followed for at least 5 years. Seven reported their photosensitivity had not changed over time, in two cases it had diminished, and in one patient the photosensitivity had disappeared.
Limitations: Retrospective study design.
Conclusion: Phototesting in children is feasible when performed in a case- and child- dependent manner. PLE was the

most prevalent diagnosis in our series followed by photosensitivity in atopic dermatitis.

PP124 - USEFULNESS OF PATCH TESTS IN CHILDREN´S HAND DERMATITIS

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Background: Hand dermatitis is not infrequent in children1, however most cases are due to irritative phenomena or atopic dermatitis. There are few reports in the literature of hand contact dermatitis in the pediatric population.
Objectives: To define the epidemiological and clinical characteristics of patients of pediatric age who underwent patch tests for hand dermatitis, also to study and interpret results of patch tests.
Methods: We performed a retrospective study of all patients of pediatric age with hand dermatitis who underwent patch test with a the baseline Spanish series in the Cutaneous Allergy Unit of the Dermatology Department of our hospital, between October 2004 and November 2009.
Results: Patch tests were performed on 130 children (0-16 years old), 35 of them had hand dermatitis (26.9%). Sex distribution was 45.7% boys and 54.3% girls. The most frequent localization was palms (25.7%). Patch tests were positive in 16 (45.7%) patients. Positive results were considered of current relevance in 12 patients (34.3%). Most frequent etiology was cosmetics (43.75%). The substance most commonly implicated was methylchloroisothiazolinone (31.25%), noting the low positivity for nickel sulphate (12.5%).
Conclusion: We highlight the usefulness of patch tests in children´s hand dermatitis. The most frequent clinically relevant allergens were those present as preservative in cosmetic products and fragances and neither nickel sulphate nor formaldehyde and releasers as observed in adults2,3.
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PP125 - FEATURES OF CLINICAL PICTURE ALLERGODERMATOSIS AT COTTON WORKERS AND CATTLE BREEDERS

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Among allergic dermatosis at cotton workers and cattle breeders the first place occupies eczema.
Skin-pathological process had basically symmetric character, at first at patients on a skin appeared a strong itch, hyperemic, infiltration, and then - small vials which were quickly opened and formed erosion, crusts, the secondary infection often joined.
At cattle breeders disease becomes aggravated during a spring and autumn hairstyle of an astrakhan sheep.
At cotton worker disease relapse is observed after processing of a cotton workers by chemicals and in the autumn during clap harvesting. At windy weather deterioration of skin-pathological process, state of health of patients is marked.
The diagnosis eczema at cotton workers and cattle breeders has been confirmed by means of the anamnesis, clinic, allergic and immunological researches (positive results allergic skin tests, hyperglobulinemia E, change of cellular immunity, an immunodeficiency, decrease gumorological immunity).
In development eczema at cotton workers the big role following adverse factors have played: the heat of air and its low humidity owing to what increases strong sweat at workers and as a result contact rural workers with the plants processed by pesticides. A considerable role conditions promoting penetration of pesticides (play scratches, cracks, the grazes, strengthened strong sweat), long (8-9) hours the working day, the big ¬ physical activities, the compelled working pose with the expressed static pressure of various muscles, lifting and manual a resock of weights.
Excessive strong sweat leads to such serious change of a functional condition of a skin as alkalization to a skin surface that promotes the expressed infringement of integrity false skin. Alkalization increase of sensitivity of a skin to exogenous stimuli and especially also promotes the chemicals applied in agriculture.

PP127 - ANTI-INFLAMMATORY PROPERTIES OF LICOCHALCONE A FROM GLYCYRRHIZA INFLATA ON HUMAN SKIN

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In the Far East licorice is a basic compound of several traditional medicines for a broad range of diseases. This study focuses on anti-inflammatory properties of licochalcone A, a retrochalcone from Glycyrrhiza inflata. The presented data demonstrate that licochalcone A is a potent inhibitor of pro-inflammatory in vitro responses of a variety of dermatologically relevant cell types, including fMLP or zymosan induced oxidative burst of granulocytes, UVB-induced PGE2 release by keratinocytes, LPS-induced PGE2 release by dermal fibroblasts, fMLP-induced LTB4 release by granulocytes, and LPS-induced IL-6/TNF-alpha secretion by monocyte-derived dendritic cells. Vehicle controlled studies with two different stress models were performed to demonstrate in vivo efficacy. Anti-inflammatory efficacy in razor shaving-induced skin irritation, and UV-induced sunburn was evaluated. In the shaving study, subjects were shaved without lubricant at test sites on the volar forearm to induce erythema. Test products were applied twice a day for three days directly after shaving and at night. Skin redness was determined on day 4 by clinical grading and by measuring the redness with a Spectrometer. In the UV-erythema study, volunteers were irriadiated on two test sites on the back. Immediately afterwards the test areas were treated either with a licochalcone A containing lotion or its vehicle. After five hours the developing erythema was evaluated by clinical grading and reflectance spectroscopy, and the lotions were applied once again. A final measurement of redness, clinical

and instrumental, was performed 24 hours after irradiation. Statistical analysis revealed highly significant anti-inflammatory efficacy of the lotion containing licochalcone A in razor shaving-induced skin irritation when compared to its vehicle. Licochalcone A also significantly reduced erythema after solar simulated irradiation.

PP128 - ANTI-OXIDATIVE PROPERTIES OF LICOCHALCONE A FROM GLYCYRRHIZA INFLATA ON HUMAN SKIN IN VITRO AND IN VIVO

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Licorice extract is frequently used in the western world as a basis for candies and as a sweetener in the food and tobacco industry. In the Far East, however, licorice is well known as basic compound of several traditional medicines for a broad range of diseases. Pharmacologic activities have been attributed to several phenolic ingredients and terpene saponins found in different Glycyrrhiza species. Licochalcone A from Glycyrrhiza inflata has previosly been shown to possess anti-bacterial, anti-parasitic, and anti-inflammatory activity.
In this study we focused on the anti-oxidative properties of licochalcone A. The production of reactive oxygen species (ROS), induced by UV irradiation and environmental stress leads to oxidative tissue damage and is one of the major causes of premature skin aging.
We examined the in vitro inhibitory effects of licochalcone A on various pro-oxidative/ pro-inflammatory reaction cascades including: fMLP or zymosan induced oxidative burst of neutrophils, UV-induced lipidperoxidation, and interference with signal transduction cascades in fibroblasts and keratinocytes.
An in vivo study with a topical formulation containing licochalcone A confirmed anti-oxidative efficacy on UV-induced ultra week photon emission.
In conclusion, these data demonstrate the potent inhibitory capability of topically applied licochalcone A against oxidative skin damage caused by ROS. This anti-oxidative activity of licochalcone A at submicromolar concentrations and the broad action profile makes it a promising candidate for dermatological and cosmetic applications, e.g. for atopic dermatitis.

PP129 - ATOPIC DERMATITIS-A RETROSPECTIVE STUDY

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Background: Atopic dermatitis is the commonest childhood dermatopathy.
Objectives: 206 children (128 boys,78 girls) with atopic dermatitis 1-5 years old,examined in the outpatient clinic of our hospital between 2007-2008, were studied.
Methods: Retrospective study of data from these patients' files. Dermal lesions and their localization, frequency of maternal breastfeeding, family-personal history of atopic disease, dermatopathy's course (exacerbations, remissions, secondary infections) were recorded.
Results: Eczematoid lesions on the extensor limp areas appeared in 147(71,4%) children, dry lichenificated lesions on pressing surfaces of elbows-popliteal fossa in 35(17%), on anterolateral cervical and ears bulbs area in 24(11,6%). Severe exacerbations appeared in 33,5% patients-mainly during cold months, microbial superinfections in 46 (22,3%). Eczema appeared during the first 3 months of life in 20,4% of cases, during the first year of life in 77,7%. Low incidence of breastfeeding was found in the majority of the patients,whereas in those exclusively breastfeeded for ~4 months or bottle-feeded with hydrolysed proteins cow milk for >3 months(35% patients), the symptoms were milder. In 54,4% of children one or both parents had allergic diseases, in 8,3% of children allergic rhinitis/asthma co-existed.
Conclusions: Atopic dermatitis appears more often in boys,it is frequently correlated with positive family history of atopic diseases and respiratory allergic diseases manifestation. Doctor-patient communication and education of patients and their parents is the key for successful treatment of the disease.
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PP130 - DIAPER DERMATITIS: ETIOLOGICAL APPROACH-TREATMENT

A. Tsiakou[*1], *E. Karachanidi*[2], *E. Michael*[1], *T. Syriopoulou*[2], *E. Kostopoulou*[3], *M. Papoula*[2], *P. Giannakopoulou*[2], *E. Fourlani*[1], *J. Mangana*[1], *A. Sidiratos*[1], *N. Karli*[2] - [1]Scientific Associate of Pediatric department, [2]Pediatric department, Kalamata General Hospital, Kalamata, [3]Scientific Associate of Pediatric department, Kalamata General Hospital, Patras, Greece

Background: Diaper dermatitis is a type of contact ekzema and one of the commonest infancy dermatopathies.
Objectives: 93 infants (29 boys,64 girls) 4-12 months old, with diaper dermatitis were retrospectively studied using the files of outpatient clinics of 2009.
Methods: Skin physical examination: inflammatory rash at gluteal region appeared in 57 infants (61,3%), erythema with exfoliation at external region in 12(13%),erythema on perigenetic region, including creases, in 23 (24,7%),corrosive "Jacquet's dermatitis" in 1(1%).
Treatment: Mild skin's cleansing, local application of hydrocortizone 1%, antifungal cream in superinfected with C.albicans cases, with satisfactory results. Precautionary recommendations were given to all the examined infants and the irritant factors were recorded.
Results: Dietary change from breastfeeding to cow milk-introduction of solid food, were 6,5% of the irritative factors, diarrhea was 9,7% of them, infrequent change of superresordent diaper was 54,8%, extensive use of talcum powder was 29%.
Conclusions: Diapery dermatitis is a non-allergic type of contact dermatitis which always improves with the proper therapy. Ascertainment and immediate removal of causing factors of dermatopathy constitute the key for its treatment.
Reference

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PP131 - THE EFFICACY OF SULFACETAMIDE 10% IN IRANIAN PATIENTS SUFFERING FROM ACNE

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Background: There are a lot of treatments for acne. Sulfur products are used to treat Acne from the time of Hippocrates, this is because of Keratolysis or irrritative effect. Sulfacetamide is very similar to PABA and compete it to make DiHydrofolic so stop making DNA temporally. The compound of Sulfur and Sufacetamide is a safe and effective drug for treating acne as in previous studies 75 % of patients were treated especially those have oily skins.
Objective: We decided to evaluate the efficacy of Sulfacetamide compound in different ethnicities of Iran.
*Materials & Method*s: In our clinical trial from all patients referred to the clinic of Tehran University we treat 56 patients of Lors, 56 of Kurds, 56 of Turks, 56 of Arabs and 56 Persians. We choose half of males and half of females. The duration of treatment was 12 weeks then we evaluate the clearness of Acne in our patients.
Results: In all of five different ethnicities the cure rate of the sum of papules, pustules, comedones and cysts were nearly 67% in Lors 69% in Kurds 69% in Turks 68% in Farses, in Arabs the cure rate was about 80% . The efficacy in Males and females were same.
Conclusions: Although the Sulfacetamide is efficient in all ethnicities of Iran included Persians, Turks, Kurds, Arabs and Lors, in Arabs it is more efficient. May be pharmacogenetic studies are needed to choose the best treatment policy for Acne in different ethnicities.

PP132 - A CASE OF FREY'S SYNDROME DUE TO FORCEPS INJURY

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A 2-year-old boy presented to the dermatology department with a history of intense erythema on the cheeks accompanied by excessive salivation during food ingestion. Initially the parents suspected a food allergy but noticed that both the erythema and salivation occur during mastication of any food item. No excessive sweating was noted on the cheeks.
On examination there was no primary dermatosis visible on the skin but when challenged with a food item he developed intense erythema on the central aspects of both cheeks, stopping at the midline of his philtrum, with hypersalivation which subsided few minutes after the ingestion. This was not accompanied by pain.
A scar was noted on the lateral aspects of the cheek in an arcuate configuration. (Figures available)
History from the parents reveal a traumatic birth of the child necessitating the use of forceps which led to the injury of the auriculotemporal branch of the trigeminal nerve. The diagnosis of Frey's syndrome was made and although he was considered for botox injections he was treated with topical glycopyrrolate 0.5% with cetamecagrol.
Frey's syndrome is also termed Auriculotemporal syndrome which can occur following damage of the auriculotemporal branch of the trigeminal nerve in parotid surgery but rarely due to forceps injury. The symptoms occur as a result of excessive regeneration of sympathetic fibers and sweating is often absent in paediatric cases. Botulinum toxin injections have been used but we have opted to have a trial with topical glycopyrrolate.

PP133 - SAPHO SYNDROME: A CASE REPORT

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Sapho syndrome (Synovitis, Acne, Pustulosis, Hyperostosis, Osteitis), is a rare disease, combining inflammatory disorders of skin and skeletal system. We present a young adolescent girl suffering from the syndrome. A 13 year old girl was admitted to our department with a 2- week history of low back pain, anterior chest wall tenderness and low grade fever. On examination she had reduced mobility of the lumbar spine, painful sternum and very severe acne. She was on clindamycin and isotretinoin p.o. for the previous 15 and 30 days respectively. Laboratory investigations showed increased signs of inflammation (leukocyte count: 18900/mcl neutrophils 80%, ESR: 105mm/h, CRP: 28mg/l). Radiographs of chest and spine were normal. Tests for rheumatic diseases were negative. Bone scintigraphy revealed increased uptake at L3 vertebra, sternum, both sternoclavicular joints and right mandible. MRI showed abnormal signal at L3, L1 vertebral bodies and the surrounding back soft tissue elements. Sternum bone marrow was negative for malignancy and osseous histology revealed osteitis. When the diagnosis of Sapho syndrome was established based in these findings , methylprednizolone (16mg p.o. tds) was added. She responded to the treatment rapidly. She received full dose methylprednizolone and clindamycin for 6 weeks each and isotretinoin p.o. for 16 weeks. During the methylprednizolone tapering, the patient experienced mild exacerbation of the back pain. Laboratory signs of inflammation and MRI findings had improved, but were still abnormal. Treatment was, therefore, continued with methotrexate, for an additional 17 months. The response to this treatment was excellent. Five years later, the patient remains in complete remission. Increased information and awareness of Sapho syndrome among physicians is needed, in order to avoid diagnostic delays as well as unnecessary investigation and invasive treatment procedures to the patients.

PP134 - Z-LINEAR ATROPHY AFTER INTRALESIONAL STEROID INJECTION FOR CHALAZIA

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Steroids can cause systemic as well as local side effects. Cutaneous changes after local corticosteroid administration include dermal atrophy, hyper or hypopigmentation and hypertricosis.
Perilesional linear atrophy is a rare, distinct and unpredictable complication after intralesional administration of corticosteroid and is probably due to lymphogen spread of the steroid suspension. Although cosmetically disturbing, this

side effect is harmful and reversible over a period of months. No treatment is recommended.
We describe the case of a 3-year-old boy who developed z-linear atrophy 1 month after the first intralesional corticosteroid injection of chalazia.

PP135 - DERMATOFIBROSARCOMA PROTUBERANS AT THE SITE OF CENTRAL VENOUS LINE: CASE REPORT

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A 9-year-old Saudi boy who presented with a soft, compressable, non-tender, dusky red colored, 3cm in diameter lump in the left supraclavicular area with scattered around deep red firm nodules and plaques. Complete excision of the tumor was performed and histopathological examination revealed a diagnosis of Dermatofibrosarcoma Protuberans (DFSP). This is the first reported DFSP occurring in a child in our country since 1994 (tumor registry incidence) whose diagnosis and proper management was delayed as it will be discussed.

PP136 - CHILDHOOD SOLITARY COLLAGENOMA

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Familial cutaneous collagenoma is an inherited connective tissue nevus which presents with asymptomatic symmetrically distributed skin nodules on the trunk or upper limbs. Here we describe a case of a 12 year old girl with collagenoma affecting the lower back.

PP137 - INDETERMINATE CELL TUMOR RESPONSIVE TO NARROWBAND ULTRAVIOLET B

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Background: Indeterminate cell histiocytosis (ICH) is a very rare cutaneous disease characterized by the presence of dendritic cells that share immunophenotypic features of both Langerhans cells (LC) (CD1a and S-100-positive) and macrophages (CD68-positive) but lack Birbeck granules.
Case report: A 4-year-old, previously healthy African American boy was referred for a 1-year history of hundreds of papules, nodules and small plaques. Histology revealed a dermal infiltrate of LC-like cells. Electron microscopy failed to highlight Birbeck granules within the cells. Immunohistochemical analysis showed that most infiltrating cells were CD1a and CD68-positive, but S100-negative. Narrowband ultraviolet B (nbUVB) was started at 2-3 sessions per week, with most lesions resolving over a 6-month course.
Discussion: This case could not be readily categorized into any of the known histiocytic disorders. It was concluded that this is the first report of S100-negative ICH. This finding supports the hypothesis that ICH actually represents a spectrum of disorders arising from a relatively heterogeneous cell population which may begin proliferating at various points in time along the differentiation pathway of LC.
Although most patients with ICH do not require treatment as the disease is often self-remitting, we decided to treat our patient because of the severely disfiguring, rapidly spreading course of the lesions. Given the patient's age and lack of systemic involvement, nbUVB was chosen for this patient because of its safety profile. The results obtained were very impressive indicating that nbUVB therapy may represent a first-line option in the treatment of ICH.

PP138 - BENIGN CEPHALIC HISTIOCYTOSIS (BCH) IN A 4 – YEARS OLD BOY – CASE REPORT

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Background: Benign cephalic histiocytosis (BCH) is a rare dermatosis in infancy / childhood period. It generally involves just the skin and is self - healing. The diagnosis is mostly set according to the clinical picture, histological and immunohistochemical findings.
Methods: A case report of 4 - years old boy is presented. The first skin lesions were noted on lateral aspects of his cheeks by his parents at the age of 4 months. Later similar skin lesions appeared also his neck, upper parts of the body and extremities.
Results: At our department of Dermatology a 4 years old boy was presented with small light brown maculo-papules. First skin lesions appeared on lateral aspects of his cheeks in the age of 4 months and in next few months similar lesions appeared also on his neck, upper trunck and upper extremities. Otherwise he was healthy.
Histopathology showed dermal infiltrate consisting mostly of histiocytes. On immunohistochemistry cells of the infiltrate were S100 and CD1a negative and on electron-microscopy no Birbeck granules were found. All these findings excluded Langerhans cell histiocytosis (LCH) and a diagnose of BCH was set. He was regulary followed by dermatologist and at the age 8 years except for some small light- brown macules on his cheecks, all other changes regressed without scaring.
Conclusions: BCH is a rare form of non Langerhans cell histiocytosis, that can be easily overlooked because of the self-resolving course. According to the literature, we expect a complete healing of the coutaneous findings, but must be aware of difficulty in differentiating between benign self-healing and aggressive forms of histiocytoses and of possible sistemic involvement. That's why we recommend a careful checking for possible sistemic findings and regular follow up of the patient till all the changes completely regress.

PP139 - MULTIPLE PLEXIFORM SCHWANNOMAS REVEALING NEUROFIBROMATOSIS TYPE 2 IN A CHILD – A NOVEL MUTATION IN THIS RARE ASSOCIATION

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Background - Neurofibromatosis type 2 (NF2) is an autosomal dominant neurocutaneous syndrome caused by mutations in the NF2 gene located on chromosome 22. Affected individuals inevitably develop multiple tumors of the central and peripheral nervous system. Plexiform schwannomas

are relatively rare, benign peripheral nerve sheath tumors, which usually arise in either the dermis or subcutaneous tissue. Previous reports have emphasized its rare association with NF2.

Case Report - A 12-year-old boy was admitted in our hospital for the investigation of right hemiparesis. He had a 4 years history of multiple small subcutaneous nodules and hyperpigmented macules on the abdomen and arms that gradually became enlarged and increased in number. Brain and medullar MRI revealed bilateral vestibular schwannomas and medullar multifocal ependymoma. He was submitted to surgery and skin biopsy. Histological examination showed a medullar non anaplastic ependymoma and cutaneous plexiform shwannoma. Audiogram and ophthalmological examination were normal. Genetic analysis revealed a novel mutation of NF2 gene, c.1114_1120delinsAA (p.Glu372fsX5), which was absent in his parents and sister. He started systemic corticosteroids and physiotherapy with symptomatic improvement. The neurologic lesions were stable one year later as revealed by MRI scan. However skin shwannomas continued to increase in size and number.

Conclusion - NF2 is inherited as an autosomal dominant trait but approximately half of the cases are caused by spontaneous mutations as in our patient. To our knowledge, this is the first NF2 case associated with this mutation. In the few cases similar to ours the presence of multiple plexiform schwannomas strongly suggested the diagnosis of NF2. However this association is rare and uncommonly reported.

PP140 - PRIMARY CUTANEOUS ANAPLASTIC LARGE CELL LYMPHOMA OF THE NASAL TIP IN A CHILD

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Background: Primary cutaneous anaplastic large cell lymphoma (C-ALCL) has been rarely described in children. We present the unusual case of a 4-year old boy who presented a rapidly developing ulcerated nodule on the nasal tip which was diagnosed as CD30-positive C-ALCL.

Case Report: A 4-year-old boy developed an ulcer on the nasal tip, covered by a necrotic schar. A skin biopsy specimen showed a neoplastic proliferation of cells disposed diffusely and in a pseudonodular pattern located in the dermis, with angiocentric disposition in some areas. Tumor cells had a median-large size, lymphoid nuclei with dispersed chromatin and abundant cytoplasms. Immunohistochemical studies showed prominent CD3 and CD30 staining by more than 75% of the atypical cells. A complete laboratory survey was normal, including bone marrow aspirates and total body CT scan. The patient was left untreated, and after 3 months of follow-up, the lesion had experienced complete spontaneous healing.

Conclusion: A conservative management is recommended in cases of C-ALCL without extracutaneous involvement. Local strategies such as excision and/or radiotherapy are justified if the solitary lesion does not spontaneously disappear after an initial period of observation. Chemotherapy may be indicated only in patients presenting with or developing extracutaneous disease. Long-term follow-up is necessary.

PP141 - PAPULAR XANTHOMA IN A CHILD: A CASE REPORT AND REVIEW OF LITRETURE

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Papular xanthoma (PX) is a very rare skin disorder. We describe a typical case of PX in a 13-month-old malay boy who presented with numerous yellowish - orange papulonodules, 2-8 mm in diameter, mainly on the face, both upper extremities, and trunk of 10 months duration. His systemic examination was normal. Serum lipid profile was normal. Histologic studies showed a diffuse monomorphous infiltrate of foamy cells in the upper dermis. The foamy cells stain positive for CD68 and negative for S-100 protein and CD1a. A diagnosis of papular xanthoma with no systemic symptom was made. The skin lesions involuted spontaneously during follow-up.

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PP142 - CONGENITAL LANGERHANS CELL HISTIOCYTOSIS WITH SYSTEMIC INVOLVEMENT

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Congenital Langerhans Cell Histiocytosis (LCH) can present as generalized papules, nodules or vesicles that resolve spontaneously without extracutaneous lesions. However, there have been cases of congenital LCH reported with multi-organ involvement. We describe a newborn with LCH involving the liver, mediastinum and lung apart from skin lesions.

An 11-day-old male neonate was referred to our department because of vesicular lesions since birth. On clinical examination he had multiple papulovesicles with hemorrhagic crusts distributed over the scalp, trunk, limbs, palms and soles. Bacterial and fungal cultures, as well as TORCH screen were unrevealing. A skin biopsy performed, revealed infiltrates of Langerhans cells positive for S-100, CD1a and Langerin, confirming the diagnosis of LCH.

Systemic evaluation, including full blood count, biochemical profile, urine osmolality, radiographic skeletal survey, was normal, whereas the abdominal ultrasound revealed three hepatic lesions. MRI and CT scans documented a mass of the right mediastinum and left pulmonary micronodular infiltration. Due to multisystem involvement and no evidence of regression, chemotherapy, including vinblastine, prednisone and 6-mercaptopurine, was initiated at two months of age. There was prompt regression of LCH infiltrates and no abnormal findings by imaging at treatment completion. At 3 years of age, the patient is asymptomatic with no signs of relapse.

LCH should be included in the differential diagnosis of vesicles in a newborn. Evaluation for systemic involvement and long-term follow –up to detect relapse are required.

PP143 - RADIO-SURGICAL TREATMENT OF LINEAR VERRUCOUS EPIDERMAL NEVUS: A CASE REPORT OF A 14-YEAR OLD GIRL

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Title: Radio-surgical treatment of linear verrucous epidermal nevus: a case report of a 14-year old girl

Introduction: Linear verrucous epidermal nevus is benign hyperplasion of epidermal cells. There is localized and systematized type of this nevus. In the localized type, which appears usually at birth, only one linear lesion is present. It may be skin colored, brown, or gray-brown. The localization may be on the head, trunk, or extremities. In the systematized type there are many linear lesions present, often showing a parallel arangement, especially on the trunk. Localized and, more commonly, systematized linear epidermal nevi may be associated with skeletal deformities and central nervous system deficiencies.

Methods: We present a case of a 14-year old female with linear reddish tumorous formation, localized in presternal region. It consists of closely set, papillomatous, verrucous papules forming well-demarcated papillomatous plaque, 6cm x 1,5cm in diameter. The lesion was present at birth and has became bigger in years. It makes a cosmetic problem to the patient, since its on the neckline.

Histopathology supported the clinical diagnosis. It shows hyperkeratosis, papillomatosis and acanthosis with elongation of the rete ridges resembling benign papilloma.

Results: After preparation of the skin and under local anesthesia, we performed radio-surgical therapy (cutting and coagulation) at medium power in two occasions. The operation procedure was followed by minimal bleeding. The lesion was dressed with antibiotic ointment and a non-adhesive bandage for a few days after the procedure.

The therapy is still not finished, but the lesions are evidently smaller in dimensions.

Discussion: Since its well known that epidermal nevi are prone to recurrences, we consider the application of topical keratolytics and retinoids as well as the possibility of repetition of radio-surgical procedure.

PP144 - RAPIDLY GROWING LYMPHOID SKIN TUMORS IN PEDIATRIC PATIENTS

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The authors present three cases of rapidly growing lymphoid skin tumors. At the time of the diagnosis all patients were under 12 years of age.

The tumors developed within 2-4 weeks.

In the first case (the tumor involved the face) the histological examination revealed primary cutaneous anaplastic large cell lymphoma (pcALCL). By the second patient the skin tumor (on the left upper arm) proved to be lymphomatoid papulosis (LyP). Both tumors together with other borderline CD30+ lesions are part of a spectrum of CD30 (Ki-1)–positive cutaneous lymphoproliferative diseases, which account for approximately 25% of cutaneous T-cell lymphoma cases. Spontaneous regression of LyP is seen almost universally, whereas regression occurs in approximately 25% of pcALCL cases. Therefore, the higher apoptotic index found in LyP compared with pcALCL is not surprising. In our patient the pcALCL resolved spontaneously, while on the boy suffering from lymphomatoid papulosis after 10 month new lesions appeared.

In the third case the histological examination of the biopsy specimen (from the left forarm) detected precursor B-cell lymphoblastic lymphoma (B-LBL). B-LBL is an uncommon high-grade neoplasm of immature B-cells, accounts for less than 10% of cases of lymphoblastic lymphoma. It is a rare subtype of childhood non-Hodgkin lymphoma (NHL). In contrast to the more common lymphoblastic lymphoma of T-cell lineage, B-LBL can be an extranodal disease, with a propensity to involve skin and bone. The bone marrow of the 2-year-old girl was not infiltrated, actually she is treated according to the NHL-BFM 95 protocol.

PP145 - RAPIDLY ENLARGING PILOMATRIXOMA OF THE EYELID

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Pilomatrixoma (also called benign calcifying epithelioma of Malherbe) is a rare, slowly growing, benign tumor originating from the matrix of the hair root, first described in 1880 by Malherbe and Chenantais. It occurs more frequently on the head and neck region of children and adolescents, often involving the eyelids.

Pilomatrixoma is often misdiagnosed clinically and the correct diagnosis can be established only after excision and histological examination.

We report an unusual presentation of a pilomatrixoma of the lower eyelid with an aggressive behavior in an 8-year-old boy and highlight on its distinctive clinical features and common misdiagnosis.

PP146 - INFANTILE MYOFIBROMATOSIS: A CASE REPORT WITH A UNIQUE CLINICAL PRESENTATION

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Introduction: Infantile myofibromatosis (IM) is a rare tumor of childhood. Herein we describe a unique case with IM that illustrates diagnostic difficulty.

Observation: A 19 month-old male infant presented to our department with subcutaneous nodules on the right lumbar region. These lesions were known to have been growing for about 5 months. At another center 9 months prior to presenting at our department, total nodule excision from the same lumbar region was performed and the pathology report stated that the nodule was infantile calcifying fibromatosis or nodular fasciitis.

On physical examination there was multiple firm, rubbery, flesh-colored nodules (7 nodules) whose size ranged from 5 to 15 mm, the bigger was ulcerated. Magnetic resonance imaging (MRI) of right lumbar region was performed, which revealed infiltrative, subcutaneous nodules in the right lumbar region without deep invasion. Histopatholigical re-study of previous nodule excised, found alternating areas of 2 di-

stinct cell populations. The lesion showed a marked proliferation of cells positive for smooth muscle actin and vimentin but negative for desmin and S-100. A final diagnosis of myofibromatosis was made. The patient was followed-up for 6 months; however, due to rapid growth of nodules, surgical treatment was suggested.
Discussion: Myofibroma (unifocal) and myofibromatosis (multifocal) are rare spindle cell neoplasms composed of myofibroblasts cells with characteristics intermediate between smooth muscle, fibroblastic, and undifferentiated cells. Lesions often undergo spontaneous remission, which may be mediated by massive apoptosis. Clinically these lesions show a predilection for the head and neck, followed by the trunk and extremities.
The case presented here represents a unique form of myofibromatosis occured on the right lumbar region with local recurrence as multiple subcutaneous nodules, the bigger was ulcerated.

PP147 - CUTANEOUS METASTASIS FROM BILATERAL ADRENAL NEUROBLASTOMA IN A 2 MONTH-OLD CHILD

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Introduction: 4S neuroblastoma with bilateral adrenal involvement is defined by small primitive tumors (stage 1 or 2) with disseminated disease restricted to the liver, skin, and/or bone marrow. Children are less than one year old. These tumors are rare and of multicentric origin. We report a case revealed by subcutaneous nodules.
Comment: A 2 month-old female infant was admitted with three subcutaneous nodules appeared a week earlier. She was the first child of healthy parents. Her prenatal, perinatal, and postnatal history was uneventful. Her growth and developmental milestones were appropriate for age. At physical examination, three firm, blue subcutaneous nodules on periumbilical region, right iliac fossa and right mammary region. Their size ranged between 3 and 10 mm. The patient presented with abdominal distension caused by hepatomegaly. Abdominal ultrasonography found two bilateral adrenal masses and hepatomegaly with heterogeneous nodules. The adrenal masses measured 77 mm on right and 35 mm on left.
Histopathological and immunohistochemical studies of one of subcutaneous nodules revealed cutaneous localization of neuroblastoma. Subcutaneous nodules regressed while other nodules appeared. Unilateral adrenalectomy was performed, and the contralateral tumor was observed. The size of the remaining tumors decreased slowly, and the clinical progress was good.
Comments: Cutaneous metastases of internal malignancies are very rare in children. In this group, neuroblastoma, leukaemia and lymphoma are the most common malignancies that may develop metastases or neoplastic infiltrates to the skin. The low incidence of these lesions and their high malignant potential are two important factors that emphasize the pivotal role the dermatologist plays in prompt diagnosis and early referral.

PP148 - FAMILIAL DYSKERATOSIS CONGENITA WITH HAEMATOLOGICAL ABNORMALITIES

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Dyskeratosis congenita (DC) is a rare inherited multisystem genodermatosis characterized by reticulate skin pigmentation (89%) nail dystrophy (88%) leukoplakia (78%) with haematological abnormalities (85%)
A 4 year old male child, 4th in birth order born of III degree consanguinous marriage presented with discolouration all over the chest and upper extremities, deformity of nails and white patch on the tongue since 8 months. He had fever since 20 days.
On examination he was anemic. He had reticulate hyperpigmentation over the chest, upper extremities palm and soles. Oral cavity showed hyperpigmentation and leukoplakia on the tongue. Nail showed dystrophy and hyperpigmented longitudinal bands. Father had DC with Fanconis anaemia and elder sister had DC with anaemia.
Investigations reveal hemoglobin of 4 gm/dl, total leukocyte count (TLC) of 2700/ cmm, platelets count of 24,000/cmm, with microcytosis and hypochromasia. Bone marrow aspiration reflected depression of all the cell lines. Cytogenetic study showed no chromosomal breakage. Skin biopsy showed basket weave orthokeratosis with hypogranulosis and thinned epidermis with pigmented basal layer. Dermis showed multiple melanophages and perivascular lymphocytic infiltrate.

PP149 - CONGENITAL BATHING TRUNK (GYANTCONGENITAL) NEVUS: DIAGNOSTICS AND TREATMENT

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Congenital bathing trunk (gyantcongenital) nevus: diagnostics and treatment. Number of children and youth patients, who address to medical institutions because of skin pigment neoplasms, increases from year to year. One of reasons for visit to a doctor is psychological and emotional problems associated with aesthetic defect; nevertheless, in some cases these masses constitute a real menace because of possible transformation to malignant melanoma.
Recently melanoma rate increases and, according to the studies, melanoma "rejuvenates", i.e. problem of differential diagnosis as well as choice of excision technology in case of skin pigment neoplasms at children and youth patients becomes actual. Congenital pigment nevuses have different sizes and can be classified by their diameter; the most risky for malignant transformation and unfavorable for aesthetics are bathing trunk nevus which sometimes can reach half of trunk skin. A serious delicacies with treatment method choice and tactics of follow-up in case of congenital nevuses are linked to a large area of affection and significant mental and physical problems for patients. In studies of children and youth patients, investigated by us, dynamic stage-by-stage excision of congenital bathing trunk nevuses and dermabrasion of moderate pigment nevuses (including "spotty" and "pilary") were used. Histological pattern was characterized by marked variety of neoplasms with melanocytic genesis variants. So, although melanoma was not seen in any of congenital bathing trunk nevuses pathological features, focuses

of dysplastic nevus of degree I-III were seen in most studies. This data demonstrates that, besides aesthetic problems, congenital bathing trunk nevuses have signs of pre-melanoma processes. It dictates necessity of active surgical treatment in case of these neoplasms; frequent trauma and focuses of chronic inflammation should be regarded as indications for excision.

PP150 - ERYTHEMA DYSCHROMICUM PERSTANS IN CHILDREN: A REPORT OF 3 CASES

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Introduction and aim: Erythema Dyschromicum Perstans (EDP) is slowly progressive, asymptomatic and an acquired pigmentation skin disorder with an unknown origin that presents with grayish or ashy macules.
The asymptomatic lesions have observed mostly at the trunk, neck and upper extremities. The disease has a benign course and tends to heal spontaneously Clofazimine and dapsone therapies has been reported to be successful.
In this report 3 cases of a rare skin disorder EDP were presented and determined under the light of literature.
Method: Three cases of EDP which clinically and histopathologically diagnosed in Pediatric Dermatology Outpatient Clinic of Dermatology Department of A.Menderes University Medical School Hospital are presented.
Results: The ages of patients diagnosed as EDP were 8, 9 and 15 respectively. No any family history or drug use has mentioned before skin lessions are observed. The common features were skin lesions as grayish or ashy macules which have seen at same regions as trunk, neck and upper extremities.
Histopathological features: Hyperkeratosis in stratum corneum, increasing of melanin pigment and a lot of melanophages are observed in biopsy investigation.
Conclusion: To verify the EDP from other pigmentation disorders is important due to various skin lesion can be seen in child patients.
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PP151 - DIPLOID/TRIPLOID MOSAICISM AND CUTANEOUS PIGMENTATION

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Introduction: Diploid/triploid mosaicism shows muscular hipotonia, body and/or facial asymmetry, mental and growth retardation, truncal obesity, clino-camptodactyly and male genital hypoplasia. Almost 50% of the patients develop early skin pigmentary anomalies.
Case report: A 12-year-old boy with no known family history of consanguinity or genetic disorders, was referred for evaluation of very light brown, linear and reticulated pigmentary patches distributed along his left hemibody. Physical examination revealed hemibody asymmetry, short height, truncal obesity, severe skeletal anomalies in hands and feet (clinodactyly, camptodactyly, syndactyly, tenar eminence hypoplasia), xerosis, thick and coarse hair, micrognathia, and normal genitalia. Past medical history was significant for growth retardation since the neonatal period, muscular hypotonia, gastroesophageal reflux, right ear hypoacusia, astigmatism, hypermetropia and learning disabilities. Laboratory studies (hormonal and metabolic) were within the normal range. Pheripheral blood karyotype was normal. Cytogenetic analysis revealed a 46 XY/69 XXY diploid / triploid mosaicism in cultured fibroblasts of the skin.
Discussion: Diploid/triploid mosaicism is a very rare chromosome abnormality. Incorporation of the second polar body into a blastomere has been proposed and in some instances confirmed as the most probable etiopathogenic mechanism. Blood karyotype is normal in most cases and another tissue must be examined to confirm the diagnosis. It is mandatory to perform a karyotype in the skin of patients with a pigmentary mosaicism and a dysmorphic phenotype in order to rule out this abnormality.

PP152 - PIEBALDISM AND NEUROFIBROMATOSIS 1: AN OCASIONAL ASSOCIATION?

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Introduction: Piebaldism is an autosomal dominant condition caused by defective cell proliferation and migration of melanocytes during embryogenesis. It is characterized by congenital depigmented white patches of skin and hair on the forehead, central chest, abdomen and limbs. Neurofibromatosis 1 (NF 1) is an autosomal dominant neurocutaneous disorder, which diagnosis is based on the criteria established by the National Institute of Health Consensus Development Conference. There have been four reports in the literature of piebaldism associated with neurofibromatosis type 1. We report e new case of piebaldism and NF 1.
Case report: A 14-year-old black girl presented since birth three depigmented patches. One located on the trunk and two on the legs, both with islands of normally pigmented and hyperpigmented macules. A tuft of white hair over the midfrontal scalp (white forelock) was also present. Multiple café au lait macules were observed at birth on the trunk and limbs. Axillary and inguinal freckling appeared during childhood. There was no history of parent's consanguinity and family history revealed inheritance for piebaldism on the great great grandmother side with 14 affected and 3 non affected members.
Discussion: Piebaldism results from mutations of the KIT gene on chromosome 4q11-q12 or deletions in the SLUG (SNA 12) gene on chromosome 8q11. NF 1 is due to an autosomal dominant mutation localized to chromosome 17 resulting in defects in neurofibromin. Whether the simultaneous occurrence of these two dominantly inherited diseases is more than a chance remains to be established.

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PP153 - HALO NEVI

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Halo nevi (HN) are defined as benign melanocytic nevi that are surrounded by an area of depigmentation. This phenomenon often indicates the beginning of involution and subsequent regression of the melanocytic nevus, a process that extends over a period of several months or years. HN is common in children and young adults, with a mean age at onset of 15 years. The etiology of HN is unknown, an autoimmune response and T lymphocytes are considered to play a key role in the progressive destruction of nevus cells. The most frequent association of HN is vitiligo, with vitiligo lesions appearing in nearby regions, as well as at other sites. HN may also be associated with some autoimmune disorders and rarely with malignant melanoma. Approaches to the management of HN in children differ from approaches in adult patients. In children and young adults, HN are common, benign, symmetrical lesions and clinical observation over time is the best policy. It is important to examine not only HN but whole skin and visible mucus membranes. Multiple lesions in children are common, so all patients with HN should be examined with Wood's filter. Children's skin is pale and multiple lesions are not necessarily seen with naked eye. However, HN may be associated with other diseases, so in children with multiple HN we recommend to do complete blood count, antinuclear antibodies, screened for thyroiditis and celiac disease. There is no need for treatment in patients with solitary HN. Halo melanoma in a child is extremely unusual and should be considered suspicious only if the halo is asymmetrical, or if the central lesion is unusual. We present the etiology, clinical presentation, possible association with malignant melanoma and approaches to the management of HN.

PP154 - THYROID ABNORMALITES IN PEDIATRIC PATIENTS WITH VITILIGO

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Background: The link between vitiligo and thyroid disease has been proved with adult patients. However, previous studies that clearly define the possible association of vitiligo and thyroid disease in children and adolescents are lacking.
Objective: The aim of our stydy to investigate the prevalence of thyroid dysfunction in children and adolescents with vitiligo and to identify any predisposing factors of this association.
Methods: The research was conducted in seventy five children and adolescence with vitiligo; 47 females (62.66%) and 28 males (37.33%). In all were studied by physical examination, thyroid ultrasonography and laboratory studies for thyroid disease.
Results: A total of 19 out of 75 patients (25.33%) had different degrees of thyroid parameter alterations. A total of 11 out of 75 patients (14.66%) had Hashimoto's thyroiditis. These patients were all affected by the non-segmental type vitiligo. The commnest onset site of vitiligo in patients with Hashimoto's thyroiditis was upper extremity, significantly more frequently compared with patients without Hashimoto's thyroiditis ($\chi 2$ $p < 0.05$). Late-onset of vitiligo in patients with thyroid dysfunction compared with patients without thyroid dysfunction (t test $p < 0.05$). There were no associations beetween thyroid dysfunction and/or Hashimoto's thyroiditis and sex, age, mean duration of vitilligo, clinical type of vitiligo and personal or family history of vitiligo and endocrine or autoimmunnity diseases.
Conclusions: Since vitiligo usually appears before the development of the thyroid disease, it may be useful to screen thyroid autoantibodies in children and adolescence with vitiligo.

PP155 - DEPIGMENTATION, REGRESSION OF CONGENITAL MELANOCYTIC NEVI IN ASSOCIATION OF HALO PHENOMENON AND OCCURING OF VITILIGO IN CHILDHOOD

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We observe congenital melanocytic nevi (CMN) periodically (every 3, 6 or 12 month) clinically and the dermatoscopic features in our department, during the last ten years.
Here we review the cases of depigmentation or regression of these nevi, and we searched for halo formation around the nevi in addition to vitiligo formation in distinctly separate locations. The regression of CMN may occure with various mechanisms, such as non specific inflammation due to a trauma, or autoimmun inflammation, desmoplasia, or without any clinical sign of inflammation or fibrosis. Family history was asked for vitiligo and autoimmune diseases. The halo phenomenon occured mainly around medium-sized or small CMN, and they were managed conservatively, mainly „wait and see" policy was decided. Excisions were only performed, if assymetry of the halo or changes of the pigmentation(color, pattern, distribution)occured. In cases of appearing simoultaneously vitiligo examination for associated autoimmun disease was performed.
In all cases a thorough search for malignant melanoma was negative. While the precise etiology of halo formation and vitiligo remains uncertain, several theories suggest that both phenomena result from an immunologic response to pigment cells. This autoimmun inflammation is able to destroy completely or partially the melanocytes of the perinevic skin, and the nevus cells of the nevus itself.
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PP156 - AN UNUSUAL PIGMENTARY DISORDER IN AN 11-YEAR-OLD GIRL

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Introduction: Pigmentary mosaicism is the term currently used to describe phenotypes with swirled streaks of hypo or hyperpigmentation along Blaschko lines, and include disorders like hypomelanosis of Ito, nevus depigmentosus and linear and whorled nevoid hypermelanosis (LWNH). They are occasionally associated with abnormal systemic features and chromosomal mosaicism.
Case Report: We describe an 11-year-old girl with skeletal and teeth anomalies, strabismus, mental retardation, and a late-onset variegate pigmentation.
There was no familiar history of pigmentary disorders.
A skin biopsy from pigmented skin in the abdomen demonstrated hyperpigmentation of the basal epidermal layer with very few scattered melanophages in the dermis.
Cytogenetic analysis of peripheral blood of the child revealed a structural X chromosomal rearrangement, secondary to a pericentric inversion present in her mother.
Discussion: This case report shares several clinical features with LWNH associated to a relevant multi-systemic involvement with an unique genetic origin when compared to previous reports.
The genetic and clinical heterogeneity of pigmentary disorders along the lines of Blaschko requires a multidisciplinary approach. The patients should be carefully screened for structural and neurological alterations, which if present should be investigated with lymphocyte and fibroblast karyotyping in order to exclude chromosomal mosaicism.

PP157 - ACANTHOSIS NIGRICANS: A LATE POSSIBLE CONSEQUENCE OF LOW BIRTH WEIGHT NEWBORN

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Introduction: Acanthosis Nigricans (AN) is a non rare dermatitis with multiple causes. It appears predominantly on the nape of the neck, in large skin folds and creases. Histologically it is a qualitatively normal hyperplasia of the epidermis linked to hyperkeratosis and papillomatosis. AN is usually associated with insulin resistance, in particular with the metabolic disorder, polycystic ovarian syndrome or it is due to an autoimmune mechanism. Other causes are paraneoplastic in origin. We are reporting a case of AN which is a late possible consequence of intrauterine growth retardation.
Observation: A 21 year old woman-in good health- presented with dark keratotic plaques on the nape of the neck, the axilla and in inframammary folds. They had appeared at the age of 13 and had been expanding progressively. The clinical suspicion of AN was confirmed by skin biopsy. She was 153cm tall for 56kg (BMI=23,9). From familial short stature, she was born premature with neonatal hypotrophy-unknown causes. No tumour syndrome was found, neither tripe palms syndrome, which could have pointed towards AN of paraneoplastic etiology. Menstrual cycles were regular and there were no signs of hyperandrogenism. Routine biological monitoring including fasting glycaemia, was normal. Oral Glucose Tolerance Test showed hyperinsulinism while IGF1 and IGF-BP3 levels were normal-same for testosterone level.
Discussion: In this case report, AN appears in conjunction with hyperinsulism, showing insulinoresistance, which may find possible consequences in the occurrence of a long term diabetic type 2. In the absence of arguments for either a hereditary origin, a hormonal pathology or an autoimmune disease, we believe that insulinoresistance is here likely a late consequence of the neonatal catch up growth which occurred during early childhood. A reduced sensitivity to insulin may also explain occurrence of hyperkeratosis in the context of chronic hyperinsulinism, aggravated during puberty.

PP158 - ASHY DERMATOSIS IN CHILDHOOD

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We report two cases of childhood ashy dermatosis in two unrelated children. A 7-year old Caucasian girl and a 9-year-old Caucasian boy both presented with multiple, asymptomatic, non erythematous, ashy-gray macules mainly on the trunk and upper arms. There was no previous intake of drugs, the general condition was unchanged. Histological examination of a skin-biopsy taken from the 9-year-old boy showed a dense, perivascular lymphocytic infiltrate and melanophages in the upper dermis. The mast cell number was normal. Serology for Borrelia burgdorferi was negative. Childhood ashy dermatosis is still considered to be idiopathic and, in contrast to the adult form, usually presents a self-limited course.

PP159 - PREVALENCE OF ORAL MUCOSA LESIONS IN PEDIATRIC PATIENTS FROM BIRTH TO 2 YEARS

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Objective: The purpose of this study was to determine the prevalence of lesions of the oral mucosa from birth to 2 years in Turkish pediatric patients .
Methods: A total of 300 infant patients from newborn to 2 years of age were evaluated from the outpatient clinics of pediatrics and dermatology departments, Fatih University Hospital, Ankara, Turkey. The mucosal lesions were documented. The data were presented as percentages and for categorical comparisons Chi-square or Fisher's Exact test were used. A p value less than 0.05 was considered statistically significant.
Results: Of the 300 infant patients, mucosa lesions were seen in only 65 (21.27%). The most common lesions were candidiasis (10.70%), Ebstein's pearls (2.68%) and geographic tongue (2.68%) in the study. The frequency of children with

mucosal alterations was higher in the group of children from 2-12 months.
Conclusion: Oral mucosa lesions can be a sign of systemic or dermatologic diseases of the infants and thereby affecting oral feeding of the infants, leading to growth retardation. Routine examination of the oral mucosa should be a part of pediatric dermatology examination.

PP160 - LOCAL HYGIENE OF PAEDIATRIC VULVOVAGINITIS

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Background: Childhood vulvovaginitis is not uncommon in general practice, paediatrics and dermatology. The child is susceptible to vulvovaginitis because of a relatively exposed vulva, a thin vaginal wall and a poor hygiene. The first line care is a good local hygiene with a non-irritating product.
Objectives: A specific product with mild surfactants, hydrating agents, the oligoelement triad Cu-Zn-Mn with healing properties and metabolised Gluco-oligosaccharide to stimulate commensal flora has been developed and tested during an open study.
Protocol: 64 young girls aged 3 to 9,8 years with light to moderate vulvovaginitis were enrolled in a European multicenter open study by dermatologists, pediatricians and pediatric nurses. The investigated product was used one to twice a day during 21 days. Clinical examination (global irritation, erythema, dryness, lichenification, oedema, excoriation symptoms, leucorrhea, pruritus, sleep disturbance, urinary burning, pain) were performed at D0 and D21. Efficacy and cosmetic acceptability of the product were evaluated by the users at Day 21.
Results: After 21 days of use, clinical examination showed significant improvement of all clinical criteria ($p<0.01$). 95 % of the Clinicians were satisfied with the product and considered it efficient in Vulvovaginitis. Strong percentages of agreement were obtained from the users. The product calmed the itches, respected genital mucous membranes and helped to protect them from irritation.
Conclusion: This study illustrated the efficacy of a product specifically formulated for intimate hygiene of young girls concerned by vulvovaginitis. The product was well tolerated and we observed a good compliance during the study.

PP161 - A NOVEL METHOD FOR THE VISUALISATION OF THE AMOUNT OF SUNSCREEN PRODUCTS APPLIED TO SKIN BY IN VIVO ATTENUATED TOTAL REFLECTION FT-IR SPECTROSCOPIC IMAGING

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There is an ongoing discussion about applied quantity of sunscreen products. For the measurement of sun protection factors (SPFs) the typical amount of product applied to skin is, according to EU and other international standards, 2 mg/cm2. Previous studies have unfortunately shown that consumers often tend to use much less than the required amount. Other SPF studies, which demonstrated different exponential and linear relationships between the SPF and the applied amount of sunscreen products, reported a decrease in SPF with a decreasing amount of sunscreen product. Not only for people with photodermatoses, photoallergies, drug-induced photosensitation or immunosuppression, but for everyone, especially for children, it is essential to have effective protection against skin damages like sunburn, photoageing and skin cancer. Fourier transform infrared (FT-IR) spectroscopic imaging with focal plane array detectors has proved a powerful technique for rapid chemical visualisation of a huge number of different samples. It offers the possibility of combining spectral and spatial information. In vivo IR imaging is an important new field of application. In this feasibility study, for the first time the application of this technique is described for the in vivo study and visualisation of different amounts of sunscreen products (0.5, 1 and 2 mg/cm2) including the typical amount a consumer normally applies on the skin. It could be clearly visualised that the amount mostly applied by consumers is only about fourth to one third of the required amount of 2 mg/cm2. With the resulting IR imaging pictures very demonstrative and convincing material is available for the first time. It can be used for patient and consumer education in order to show and assure them how important the right amount of applied sunscreen product is.

PP162 - FACIAL CHANGE IN HEMOGLOBIN E/BETA THALASSEMIC PEDIATRIC PATIENTS

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Background: Thalassemic facies (or facial changes) are major consequences of over erythropoiesis and causes of psychiatric problems.
Objective: To study the rate of facial changes in a year among severe and non-severe hemoglobin E/beta thalassemic patients.
Patients and methods: We measured: head circumference above the eyebrows over the occiput; head circumference at ears' upper attachment points to the lowest part of the nasal septum and over the occiput; the distance between the most prominent part of the zygoma; and, the distance between the lowest part of both ears to the jaw's lower rim. The study was performed in 45 severe and 18 non-severe hemoglobin E/beta thalassemic patients and 30 control subjects.
Results: The respective male to female ratio and age (years) of the severe and non-severe thalassemic patients and control subjects was: 24 to 21 and 11.86±4.44; 6 to 12 and 11.25±4.01; and, 12 to 18 and 11.65±1.41. The mean steady-state hemoglobin(g/dL) and packed-red cell transfusion (mL/kg/year) for the severe vs. non-severe group was 6.81±0.90 and 8.42±0.71 vs. 94.30±25.36 and 70.70±25.54. A significant difference was found in: (1) the mean zygoma's distance (cm) between the severe group and controls [-0.49 (95 % CI: -0.80, -0.18)] and between the non-severe group and controls [- 0.45 (95% CI: -0.85, - 0.04)]. The distance between the lowest part of both ears in the severe group and controls was significantly different [-0.37 (95% CI : -0.73, -0.01)]. None of the differences in means measured between the severe and non-severe groups were statistically significant.
Conclusions: Since all of the patients with Hb < 9 g/dL were

given 10 mL/ kg of leukocyte-poor, packed-red cells to suppress over erythropoiesis, the findings indicate that regular blood transfusions make no significant difference to the progress of facial changes among sufferers of severe and non-severe hemoglobin E/beta thalassemia.

PP163 - DYNAMICS OF RETROGRESS OF SYPHILIDES FOR PATIENTS THE SECONDARY RECURRENT SYPHILIS, TREATED EXTENCILLIN IN COMBINATION WITH IMMUNOMODULIN AND A-TOCOPHEROL ACETATE

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The purpose of our probes was the comparative assessment of dynamics of clinical developments of a secondary recurrent syphilis for patients, treated with extencillin in a combination and w/o immunomodulin and vit E. Probes were spent in two representative bunches of patients (on 24 patients) with a secondary recurrent syphilis. Among them there were 26 men and 22 women.

Specific therapy implemented extencillin. As for sick of a syphilis decrease of activity of ferments of an antioxidative guard occurs assigned vit E on 0.2 g 2 times in day in flow of all course of specific therapy. The primary group of patients has received vit E and immunomodulin (on 1,0 i/m daily N 10), and matching bunch - against extencillin only immunomodulin.

Analysis of dynamics of times of retrogress of syphilides for patients has displayed the following. Clinical developments roseola emptyings transited in time from 3 till 10 days, on the average 5,8 days for patients of a primary group, and in times from 6 till 15 days on the average 8 days - for patients of bunch of matching.

Comparing outcomes of rate regress syphilides for sick of the secondary recurrent syphilis, receiving treatment extencillin in a combination with immunomodulin and vit E and without last it is visible, that antibacterial therapy with an immunocorrection and an anti-oxidising agent significantly accelerates retrogress of syphilides.

These data bear that the combination of an immunocorrection to antioxidatic therapy for sick of a syphilis against extencillin is pathogenetically reasonable, as for the yielded class of patients clinical developments of the syphilis, the concomitant pathology, is social-personal characteristics of patients can be labels of the secondary immunologic unsufficiency, requiring purpose of immunomodulating factors.

PP164 - IMMUNOLOGIC MONITORING AT TREATMENT OF EARLY FORMS SYPHILIS WITH EXTENCILLIN IN THE COMBINATION WITH IMMUNOMODULLIN AND A-TOCOPHEROL ACETATE

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The purpose of the present probe was carrying out of immunologic monitoring for sick of early forms of the syphilis, receiving specific therapy with extencillin (as requested) in a combination to the immunomodulating factor immunomodulin and an anti-oxidising agent a-tocopherol acetate.

It has been treated by 95 sick syphilis among which one for 45 troubleshot secondary -recurrent, for 50 - a blended early syphilis. An age of patients - from 18 till 44 years. Men - 58, women - 37. Concomitant diseases have been troubleshot for 42 patients. Among observable 6 - used junks, 26 - misused with alcoholic beverages. Clinical developments and serological a pattern of blood for patients were typical for the syphilis modern trend. An assessment(evaluation) of the immune status fabricated with usage of monoclones. Patients inspected before treatment. In a reference group inspected 15 practically healthy faces of both sexes at the age from 20 till 40 years.

In clinical bunches of patients of value CD-4 of lymphocytes bore ABT insufficient “helper” activity of T-cages. After the conducted treatment for the patients receiving against extencillin, immunomodulin and a-tocopherol acetate, the relative indexes CD-3 have considerably increased. Absolute index CD-4 of cages was above for the patients receiving complex nonspecific therapy. Application extencillin in a combination to complex nonspecific therapy contributed in the expressed correction of indexes of phagocytic activity of neutrophils.

Thus, the T-cellular immunologic unsufficiency caused by decrease of a pool of T-cages and their decrease “helper” of activity, and also unsufficiency of a nonspecific reactivity was characteristic of peripheric blood sick of lueses the secondary recurrent and blended early.

PP165 - EPIGEN-INTIM IN TREATMENT OF UROGENITAL HERPES

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We applied a drug to treatment the Epigen-intim which one active agent is activated glicerrisine acid which is one of operating ingredients of the radical of a licorice. It has antiviral and antiinflammatory operation. The Epigen-intim disinfectant, antiinflammatory, antiherpetic a resource in the form of a spray. One of singularities glicerrisine acids is the unique constitution of the formula yielding affinity to histic receptors of steroid hormones, direct anti-virus an effect has.

We have applied an Epigen-intim for 11 patients with recidivous genital herpes. Among the inspected patients would be women and 5 men. The age of patients varied from 18 till 52 years, and faces young and an average age (from 21 till 40 years) have compounded overwhelming majority - 78%. The prescription of disease compounded from 2 month till 4 years. The Epigen-intim was put on staggered districts of 5-6 times in day within 7 days. Treatment started during backsets, including: in a prodromal stage for 4 patients, in the first day of a backset for 4 patients, for the second day - for 3 patients. Monitoring implemented from the second backset till the eighth day and further with a spacing in 1 month still half a year. Outcomes of treatment depended on a phase in which one therapy has been started: it that was more effective than earlier Epigen-intim application started. Effectiveness of treatment was high for 4 patients when emptyings aborted within 1-2 days, accurate effectiveness was watched for 6 patients, thus duration of disease has contracted in 2 ti-

mes. For one sick effect it was not marked. Thus all patients marked the expressed anaesthetising and antipruritic effect. Thus, external therapy by the Epigen-intim of backsets of urogenital herpes is effective and friend in application by a method of cupping of exacerbations.

PP166 - PARTICULAR CASE OF ACTINOMYCOSIS

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The patient A. 1962 years of birth, worked as the aid-woman. Has entered the Samarkand Regional dermatovenerologic dispensary 4.05.2004. It is directional from clinic of the Samarkand Medical Institute where was on hospitalisation in therapeutic compartment concerning the diagnosis a pyelonephritis. As a halter for hospitalisation in dermal compartment of a dispensary the deterioration of a general status bound to advance of skin-pathological process has served. At entry a moderately severe status, consciousness-clear, a rule in sand bed the compelled. A constitution asthenic, a skin pale, dry, with the under turgor. Inguinal, axillary, cubital lymphonoduses at a palpation are increased before the bean dimensions, densely-elastic consistencies, with an ambient tissue are not soldered, slightly painfull. Dermo-pathological process of wide-spread nature with a lesion of a skin of a mean third of dextral forearm, a flank surface of the dextral hip where on hyperemic a hum noise there were knot holes from which one were secreted grey-yellow purulent chipses masses. From them druses aciynomyces are laboratory evolved. On a belly heel-nodulose educations were determined subcutaneous woody tightness. At ultrasonic of an internals in both nephroses and submucoses a stomach knots in the dimension 2x2 are revealed see
The diagnosis: a gummoz-knotty actinomycosis.
It has been assigned: Penicillinum on 500 000 UN 4 times a day, Ftivazidum on 1 tab. 2 time in day, solution of a potassium of iodine of 2% on 4 drips 2 times a day. Local: 10% Unguentum Ichthyoli. For 3rd day of treatment the common status of the patient was sharply aggravated, discovered a stomachal bleeding in this connection she has been translated in intensive care unit where has in a day deceased.

PP167 - MICRONIZED PARTICLES IN A SUNSCREEN FORMULATION ARE NOT DETECTED BEYOND THE STRATUM CORNEUM IN ADULTS AND INFANTS

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Background: Exposure to natural and simulated ultraviolet light (UV) has been associated with numerous adverse consequences, most importantly, an increased risk of skin cancer. Emerging evidence suggests that protecting from the harm of UV light early and throughout life is an important part of the approach to minimizing total risk of exposure. Physical/barrier sunscreen filters have been effectively used for baby- and child-recommended sun protection products for decades. Although there is considerable evidence and history of safe use of products containing ZnO and TiO2 of various particle sizes, little data exists regarding the absorption of nano-sized particles through human skin. Objective: To evaluate the absorption characteristics of a sunscreen containing ZnO/TiO2.
Methods: Two non-invasive methods: Confocal Laser Scanning Microscopy (CLSM) in reflectance mode and Confocal Raman Microspectroscopy (CRM) were used to evaluate the absorption characteristics of a sunscreen formulation containing micronized ZnO and TiO2 in 12 adults and 10 infants at baseline and after topical application.
Results: CLSM images show that the particles concentrate on the top of the stratum corneum and at the microrelief lines in both adults and infants after single application. CRM spectra was used to construct concentration profiles of TiO2 through the stratum corneum and viable epidermis and these did not show penetration of the particles beyond the stratum corneum of adults or infants.
Conclusion: In agreement with the preponderance of previous reports on adult skin, no penetration of micronized particles of ZnO/TiO2 was detected beyond the stratum corneum in the infants evaluated.

PP168 - UNUSUAL SKIN CHANGES IN 4-YEAR-OLD SCID PATIENT

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Background: Severe combined immunodeficiency (SCID) is genetically heterogeneous syndrome of recurrent infections, diarrhea, dermatitis and failure to thrive, leading to severe T and B cells dysfunction. Omenn syndrome is form of SCID additionally characterized by erythrodermia, lymphadenopathy, hepatosplenomegaly, eosinophilia and elevated IgE.
Objectives and Methods: In primary immunodeficiency registry of Department of Immunology there are 39 SCID patients
Results: 4-year-old girl with bronchopneumonia and maculo-papulo-scarring eruptions on extremities and face was admitted to Immunology Department. By 18 months she suffered from recurrent pneumonias, oral thrushes and failure to thrive. Concomitantly unusual skin changes appeared on extremities, next on face. Blood tests showed lymphopenia, no eosinophilia. elevated IgG, decreased IgA, IgM and normal IgE; IgG subclasses and cellular immunity impaired; no maternal engraftment, radiosensitivity at low level. HCMV and Aspergillus infections were detected. In BMA no signs of malignancy.
Skin examination showed histiocytes and lymphocytes infiltration. Diagnosis was T-B-NK+SCID. Mutations in RAG1/2, IL7RA were excluded. Treatment consisting of antibacterial, antiviral, antifungal therapy, IVIG and immunosuppressant (Cs-A and steroids) did not cause skin lesions healing. In March 2007 the girl underwent MUD PBPC. In early outcome GVHD II in skin (+20day), reactivation of HCMV (+27day), complete chimerism (+39day) were ascertained. During last follow-up she was in good clinical condition, only mild scars on skin.
Conclusions: Erythrodermia is typical for Omenn syndrome. Our patient presented unusual skin lesions. Skin changes are

of great importance in recognition of SCID patient, especially Omenn syndrome.

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PP169 - CUTANEOUS PATHOLOGY IN SUB-SAHARAN CHILDREN. TWO YEARS EXPERIENCE IN A TERTIARY HOSPITAL IN A MEDITERRANEAN AREA

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Introduction: The increase in the immigrant population in Europe with possible differences in the epidemiology of some skin diseases may challenge our clinical activity.

Objective: 1) To know the epidemiology of sub-Saharan children attended in our area. 2) To describe the kind and frequency of the different cutaneous pathology. 3) To analyze the frequency, presentation form and consultation way.

Material and methods: All the children under 14 years old, born in a sub-Saharan country or with at least one parent born in those countries were asked for epidemiological information. Data from clinical history and from dermatological exploration were collected over a two-year period.

Results: 27 sub-saharan children were studied from a population of 548 immigrant children seen between 2006 and 2007 (4.9% of all the immigrant children). The average age was 57.9 months (51.9 % boys and 48.1 % girls). 77.8 % of them only consulted once. The origin of the children was: Nigeria (5), Cameroon (3), Ethiopia (1), Guinea (6), Senegal (3), Congo (3), Mali (2) and Guinea Bissau (1). 16 (59.2%) were children of immigrants whereas 9 (33.3%) were immigrants and 1 child was adopted.

The most frequent entity reported as a single diagnosis was atopic dermatitis (30.8%) followed by molluscum contagiosum (7.7%) and ectoparasithosis diseases. When taken as a whole, the main diagnosis group was infectious (37% of the visits).

Discussion: We would like to point the following aspects:

1) Sub-Saharan immigrants are not a frequent population in our Departement

2) The average age was 5 years old. Most of them were children of immigrants.

3) Atopic dermatitis was the most frequent diagnosis, but infectious diseases was the main diagnosis category.

3) No exotic or tropical disease was observed in this immigrant population.

PP170 - RIGA-FEDE DISEASE : FROM SEVEN MONTHS TO SEVEN YEARS

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Introduction: Riga-Fede disease is a rare sublingual traumatic ulceration. It results from repetitive trauma of the tongue caused by lower incisors.

We report two cases of Riga-Fede disease occurring in different situations.

Observation: A seven month-old girl was referred to the neonatal ICU for iterative convulsions. She was hypotonic, blind, and deaf. Cerebral CT scan showed dural hematoma and meningeal haemorrhage, resulting from a shaken baby syndrome. Within two weeks, she developed a large ulceration on the ventral side of her tongue facing her two recently erupted lower incisors. As a consequence of brain injury, she had fly-catcher's tongue, swallowing impairment and trismus. Riga-Fede disease associated with encephalopathy was diagnosed. Teeth extraction led to complete healing. A healthy seven year old boy was suffering from a relapsing and prolonged painful rounded ulceration on the tip of his tongue. Viral infection and autoimmune bullous disease had been previously suspected and various treatments had been tried unsuccessfully. Teeth examination led to a diagnosis of Riga-Fede disease. His lower incisors were irregularly spaced and rotated with their lateral edge facing the tongue. An orthodontic treatment was proposed.

Discussion: First discribed in 1881 by Antonio Riga, Riga-Fede disease is more frequent in the neonatal period. Its early recognition is important to avoid misdiagnosis and ineffective treatments. Denutrition is the major complication. Treatment is orthodontic.

Conclusion: Riga-Fede disease can be observed in very different situations. It is self evident in infants with abnormal lingual movements but can be more puzzling in otherwise healthy older children.

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PP171 - 6MP WITHDRAWAL FACIAL ERUPTION

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Background: 6-mercaptopurine (6MP) and methotrexate (MTX) are used as maintenance therapy for acute lymphoblastic leukemia (ALL) in children. We report four cases of eruptions with the same clinical, topographical and chronological characteristics, probably related to treatment.

Case reports: Four six-year-old children with pre-B ALL in clinical remission were referred from 2006 to 2009. They were having maintenance chemotherapy involving 6-MP and MTX and were taking thrice a week prophylactic cotrimoxazole for 24 months. Physical examination revealed erythematous papules and pustules of the face, upper chest and forearms with photosensitivity, occurring on cessation of treatment or few weeks after. Histopathologic examination showed interface dermatitis with few necrosis bodies and folliculitis. There was no evidence for systemic lupus erythematosus. Topical corticosteroids were allowed no improvement. Complete recovery was achieved with topical metronidazole within a month.

Discussion: This distinctive eruption may be due to withdrawal of combination of 6-MP and MTX. Its distribution suggests a photosensitive element but has never been asserted by photobiological exploration. MTX is a known photosensitizer; 6-MP is converted into 6-thioguanine that acts as a chromophore for UVA. Moreover, in view of this rosacea-like eruption gradually cleared with topical metronidazole,

demodicidosis should be suspected since it has been well described as an opportunistic infection of the skin in immunocompromised children.
Conclusion: Distinctive rosacea-like eruptions with photosensitivity occur following withdrawal of 6-MP and MTX used as maintenance therapy for children's ALL on cessation or after a long period-treatment, when immunosuppression isn't the deepest. Their pathogenesis remains unclear.
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PP172 - TOPICAL CORTICOSTEROID PHOBIA IN ATOPIC DERMATITIS: A STUDY OF PREVALENCE AND ASSOCIATED FACTORS

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Introduction: Atopic Dermatitis is a common chronic cutaneous disease, which has an important impact on quality of life. Topical corticosteroids are the first line therapy and are very efficient. Many therapeutic failures seem to be induced by a bad adherence to medication due to corticosteroid phobia or fear to use topical corticosteroid. The aim of this study is to analyze the prevalence and the associated factors of corticosteroid phobia in atopic dermatitis, and its impact on adherence to treatment.
Method: We realized a prospective, multicentric study from February to May 2009 in 5 centers in France. 9 from hospital and 53 private dermatologists participated in the study. A 51 items questionnaire was developed after a qualitative and explorative analysis according to the focus group methodology.
This questionnaire has been administered to each patient with atopic dermatitis or to their parents attending consecutively the 5 dermatology outpatients departments and private dermatologist centers. The data were analyzed with univariate and multivariate analysis.
Results: 208 patients were enrolled and answered exhaustively to the questionnaire. 80.2% of the patients admitted to have fears about topical corticosteroid, and the intensity of the fears reached an average of 4.2/10 (+/-2.78). Only 10% admitted to be confident in using topical corticosteroids. 36% of the patients admitted non adherence to treatment.
The univariate analysis found a correlation between corticosteroid phobia and the youth of the child ($p<0.05$), a lack of clarity about how to use the treatment ($p<0.05$), a lack of trust in their practitioner ($p<0.001$), discrepancies in the informations given by the practitioners ($p<0.05$), and the past experience of an adverse side effect ($p<0.05$). Furthermore corticosteroid phobia was linked with a worse adherence to treatment ($p<0.001$), and to treat oneself the later or the shorter ($p<0.001$). Corticosteroid phobia wasn't associated with the characteristics of the atopic dermatitis.
The multivariate analysis confirmed a correlation between these variables and corticosteroid phobia.
Discussion: This study confirms the important prevalence of the corticosteroid phobia among patients with atopic dermatitis or their parents in France, and the impact of this fear on treatment adherence.
The fears were varying in their intensity and in their object. The corticosteroid phobia isn't linked with the characteristics of the atopic dermatitis but rather with the quality of the information delivered by the practitioner to the patient, and the role of the doctors and chemists appear to be a major element in the origin of this fear.
In addition we confirm the important impact of the corticosteroid phobia on the adherence to treatment. Thus corticosteroid phobia could be a cause of therapeutic failure in atopic dermatitis.
Conclusion: Corticosteroid phobia exists and is common among patient with atopic dermatitis. Its prevalence, its impact on adherence, and the fact that it concerns all kind of patients are arguments to search for those fears in clinical practice.

PP173 - ALLERGEN MICROARRAYS: A NOVEL TOOL FOR HIGH-RESOLUTION IGE PROFILING IN INFANTS AND CHILDREN WITH ATOPIC DERMATITIS

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Background: Proteomic microarray technology has recently been introduced into clinical allergology, but its applicability in children with atopic dermatitis (AD) has not been investigated so far.
Objective: To evaluate the utility of an allergen and staphylococcal superantigen microarray in the diagnostic workup of children with AD and to correlate the obtained IgE recognition patterns with clinical patient characteristics.
Methods: Specific IgE antibodies (sIgE) against a repertoire of 96 microarrayed allergen components and 10 staphylococcal superantigens were determined in serum samples of infants and children with AD. Associations of sensitization patterns with disease severity, atopic comorbidity, family history, impetigo and total serum IgE were analyzed.
Results: 140 patients (55 girls, 85 boys, mean age 74 + 53 months) with mild to severe AD (mean SCORAD 36 + 29) entered the present study. At a mean total IgE of 1528 + 3820 kU/l, microarray analysis revealed a range of 0 to 33 sensitizations against inhalant and nutritive allergen components per individual patient. 24 patients revealed sIgE against at least one staphylococcal superantigen.
The total number of allergen-specific sensitizations was significantly higher in patients with severe AD as compared to only mildly or moderately affected children, whereas superantigen-specific IgE antibodies were not differentially expressed in patients with different disease courses. Cluster analysis disclosed a panel of 10 allergen components that were associated with both disease severity and the occurrence of impetigo.
Conclusion: The current study provides evidence that allergen microarrays represent a promising tool for component-

resolved diagnosis in children with AD. However, further large-scale studies in unselected patient populations are warranted before the allergen microarray's introduction into daily clinical practice.

PP174 - ACTUAL PROBLEM DIAGNOSTIC - DYSPLASTIC NAEVI IN THE CHILDHOOD

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The melanocytes are cells located in the skin and synthetizing pigment melanin. Melanocytic tumors include a large variety of benign and malignant neoplasms. From a clinical and morpological public health of view, the naevus and melanoma are the most important. Primary prevention and differential diagnosis the naevus and melanoma are considered the most promising approach to a reduction of melanoma mortality. Cases of melanoma in children younger than 10 year have been recorded. Characteristically, lesions first appear around the time of puberty, it is unlikely that an individual has the familial melanoma/dysplastic naevi trait. Dysplastic naevi may progress to malignant melanova, this lesions occur as sporadic naevus and in a familial setting /1, 3/. The incidence of melanoma developing in a given dysplastic naevus has been estimated /at 1: 3000 per year/. Patients with greater numbers of dysplastic naevus are at greater risk for developing of melanoma / 4 /. And risk of melanoma was greater for children, who tend to make this naevi with high-grade histological atypia.

The major histopathologic criteria include architectural and cytologic features: mild to moderate atypia melanocytes, stromal reactions and elongated rete ridge. There is lack of consensus regarding the histologic classification and our clinico-histologic classification, it is included the general principles two classifications of melanocytic skin tumours and ease of coding classificatins were needed according to histological type, clinical tipe with degree of malignancy / 5 /. Melanoma risk has been associated with the degree of architectural and atypia melanocytes. It was concluded that the risk was greater for children who tend to make naevi with high – grade histological atypia. The clinical and histological differential diagnosis of dysplastic naevi includes melanoma in situ, minimal melanoma / 6 / and Reed naevus.

Currently, histologic and clinical changes of dysplastic naevus are often observed at the peripheries of primary melanomas not associated with naevi and such finding have been interpreted as representing «precursor» dysplastic naevi /672/.

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