ORAL PRESENTATIONS

OP001 - INTENSIVE CARE FOR NEONATAL SKIN DISORDERS

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Background: Neonatal skin disorders may range from benign and self-limited to life-threatening and fatal diseases, in some cases associated with other organs involvement or syndromes.
Objectives: To access the clinical features of different skin disorders in a series of newborns, at a level III Neonatal Intensive Care Unit (NICU).
Methods: We conducted a review of all newborns diagnosed with neonatal skin disorder that required intensive care over the last 13 years.
Results: 25 neonates were identified, with a male/female ratio=1:1.5. According to the cutaneous presentation at admission, skin disorders were divided into 5 major groups: blistering diseases - 7, ichthyoses - 5, aplasia cutis - 5, vascular lesions - 4, and other disorders - 4. Consanguinity was present in 3. Ten (40%) were preterm. The median gestational age was 37 weeks (27-41) and the median birthweight of 2324g (730-3650), and 7 (28%) disclosed intrauterine growth restriction. Three chromosomal disorders (13 trisomy), one LMNA mutation (restrictive dermopathy), one connexin-26 mutation (KID syndrome), one NEMO mutation (incontinentia pigmenti) and 4 other dysmorphic associations were observed. Eight (36%) neonates needed mechanical ventilation. There were 5 (20%) fatal cases. The median NICU stay was 13 (2-112) days. After NICU discharge, 12 neonates remained in the Department of Pediatrics for a median period of 16 days (5-221).
Conclusions: Our experience reveals that some skin disorders of the newborn need intensive care, some are associated with other major anomalies or genetic conditions and fatality is the hazardous outcome in a few cases. These infants impose a multidisciplinary approach and a long term follow-up of the survivors.

OP002 - CUTANEOUS MANIFESTATION OF HIV INFECTED CHILDREN:ONE CENTER EXPERIENCE

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Cutaneous manifestation are common in HIV infected children.In some studies,prevalence up to 100% had been quoted.In Paediatric Institute,Hospital Kuala lumpur, a study done by Rosnah et al in 2000 found that 80% of children infected with HIV had some form of cutaneous disease.
Objectives of this study is to look at prevalence of cutaneous manifestation in children infected by HIV and type of cutaneous involvement.
This is a cross sectional study done in HIV infected children attending our clinic from June 2008 to May 2009.
There were 123 children on follow-up with 1-2 new cases per month of HIV infected children.The prevalence of cutaneous symptoms in HIV infected children was about 60%.The most common skin disorder was pruritic papular eruption(PPE) followed by drug rash and infectious causes. For drug eruption, the commonest was macular papular rash followed by urticarial type and one patient with Steven Johnson Syndrome.All of the drug eruption was secondary to Non nucleoside reverse transcriptase inhibitor (NNRTI).The infecting agents causing cutaneous disorder is herpes that presents as shingles and fungal infections. Most common fungi is still candida causing oral/esophageal candidiasis with one patient having pyoderma gangrenosum secondary to aspergillosis another child with molluscum like lesion.All children with fungal infection had CD4 less than 15%.
Cutaneous manifestation in HIV infected children is still common with non infectious manifestation of PPE topping the list.
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3) Rosnah et al.: Mucocutaneous manifestation of pediatric HIV/Aids. Jurnal Dermatologi Malaysia 2000;vol 13:20-24.

OP003 - DILEMMA OF MANAGING PEMPHIGUS IN CHILDREN

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Corticostaroids have remained the mainstay of management of childhood pemphigus. High dose oral corticosteroid required for control of disease activity is associated with multiple long term serious complications. Other immunosuppressive agents and immunomodulators are generally not effective alone and are used as adjuvants to corticosteroids. They are either costly in a resource poor setting or causes significant long term side effects.
Between January 1994 and December 2007, 28 children [<15 years, 19 pemphigus vulgaris (PV) and 9 pemphigus foliaceous (PF)] with pemphigus were seen. All of them were treated with dexamethasone pulse (DP) therapy (100 mg intravenous infusion in 3 consecutive days every month). Cyclophosphamide was given at a dose of 25 mg/ day orally in the intervening period. Pulse therapy has been divided in the following phases: phase 1- till 6 months after achieving clinical remission when monthly pulses are given along with oral cyclophosphamide; stage 2- till 12 months after stage 1 when only oral cyclophosphamide is given; and stage 3- treatment free follow up while the patient is in remission.
Twenty four patients are in stage 3, 2 patients each are in stage 1 and 2. In those patients who are in phase 2 or 3, average number of pulses required to attain lesion free status in PV was 11 (range 7- 16) and 15 (range 9-27) in PF. In general, number of pulses required was directly proportional to the severity of disease to start with. Average length of follow- up from the diagnosis was 6.7 years. Of 24 patients in phase 3, 15 are of PV and 9 are of PF. Of them, 13 have completed a follow- up of 5 years and 11 are in 2-5 years period. Only 1 patient had minimal relapse while in phase 3, which was

mild and controlled with dapsone and topical steroids. No significant side effects were observed in any of the patients. In conclusion, DP is safe and effective treatment option in childhood pemphigus.

OP004 - DISSEMINATED CONGENITAL COMEDONES

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Background: Comedones in a newborn are an infrequent finding. We present a child with extensive congenital comedones on his face, neck, upper trunk and proximal upper limbs.
Case report: A newborn had asymptomatic, widespread, open comedones located on the upper trunk, face and upper limbs were seen, with a bilateral and symmetric distribution skin comedones since birth. The scalp, palms, soles and mucous membranes were spared. Analytical and hormonal testing was normal. A skin biopsy revealed closely set, dilated follicular infundibula with prominent orthokeratotic pluggings. No treatment was instituted. The lesions persisted unchanged, but by the age of 2 years, some lesions have started to disappear, leaving minimal or no scarring.
Conclusion: Our patient's features do not fit with any of the reported diseases which present extensive comedo appearing at birth or during childhood, including nevus comedonicus, familial dyskeratotic comedones, idiopathic disseminated comedones, childhood flexural comedones, and acne neonatorum.

OP005 - INFLIXIMAB INFUSION FOR NETHERTON SYNDROME: SUSTAINED CLINICAL IMPROVEMENT CORRELATED WITH A REDUCTION OF TSLP (THYMIC STROMAL LYMPHOPOIETIN) LEVELS IN SKIN

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Background: Netherton syndrome (NS) is caused by mutations in SPINK5, a gene encoding the protease inhibitor lymphoepithelial Kazal-type–related inhibitor (LEKTI). It has been shown that the pro-Th2 cytokine thymic stromal lymphopoietin (TSLP) and pro-inflammatory cytokines such as TNF-alpha are overexpressed in NS epidermis. We report a case of NS with high TSLP and TNF alpha cutaneous expression, who presented a dramatic clinical improvement under infliximab therapy.
Case report: A 25-year old woman with NS presented with recurrent severe inflammatory flares unresponsive to traditional therapy. Infliximab infusions (5mg/kg) were given (week 0-2-6 and every other month). A dramatic improvement was observed after the second treatment cycle, and after one year of therapy, the skin was nearly clear of inflammatory lesions.
TSLP expression was assessed by ELISA and quantitative RT-PCR in skin biopsies obtained from inflammatory and non inflammatory lesions before and after infliximab therapy. ELISA revealed an elevated TSLP production in inflammatory skin lesions before and after infliximab therapy. After one year of anti-TNF, a reduction by 4 of TSLP production was observed both in inflammatory and non inflammatory skin lesions. Quantitative RT-PCR performed on total RNA isolated from skin biopsies gave similar results for TSLP and revealed a strong expression of TNF-alpha in both lesional and non-lesional skin that was not modulated by infliximab therapy.
Conclusion: Our data suggest that TNF may play a crucial role in NS and in the inflammatory cascade driven by TSLP, and that an anti-TNF should be considered as a valuable therapeutic approach for NS.
Reference
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OP006 - CONGENITAL PARRY-ROMBERG SYNDROME?

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Case report: A male neonate born at tem to a 36-year-old mother was examined shortly after birth because of congenital left hemifacial lipoatrophy. He had wrinkled and erythematous skin with prominent veins and midline limitation. He also had three annular erythematous macules without atrophy on his left thigh, right buttock and left knee. He had no visceral involvement. Serologic tests for viruses, Borrelia burgdorferi and syphilis were negative. Anti-nuclear antibodies were absent in the infant but positive (1/320) in his mother. Abdominal, cardiac and transfontanellar cerebral ultrasonography, blood tests and karyotype were normal. Annular macules had resolved by 2 months without scarring. No skin biopsy was performed. A facial MRA showed left sided lipoatrophy, and asymmetry of the underlying bone structures, but no arteriovenous malformation or blood flow asymmetry. Hemifacial atrophy remained stable for two years. Psychomotor development was normal.
Discussion: Several diagnoses were considered. Parry Romberg syndrome (PRS) was is a rare condition of unknown origin characterized by unilateral and progressive atrophy of facial skin, subcutis and underlying bony structures. Although it is usually acquired, congenital occurrence has previously been reported. In our patient, intrauterine infections, chromosomal mosaicism and neonatal lupus erythematosus were also considered but no evidence was found. Wiedemann-Rautenstrauch syndrome is a progeroid syndrom with absent subcutaneous fat but the absence of associated anomalies stood against this diagnosis. Annular erythematous rash was suggestive of neonatal lupus but no further evidence was found.

OP007 - NEONATAL PURPURA FULMINANS

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We present a newborn with the typical features of purpura fulminans due to congenital homozygous protein C- defi-

ciency. Congenital homozygous or compound heterozygous protein C- deficiency is an important differential diagnosis in neonatal purpura fulminans. Patients develop disseminated intravascular coagulation as a consequence of accelerated microvascular thrombogenesis. Cerebral infarction and following cerebral hemorrhage as well as intravitreous hemorrhage are typical complications. Diagnosis is based on the characteristic clinical presentation and decreased protein C-plasma concentrations. Without appropriate therapy the outcome is fatal in most cases.

OP008 - CD30+ LARGE ANAPLASTIC CELL LYMPHOMA (ALCL) PRESENTING AS FEBRILE ULCERONECROTIC MUCHA HABERMANN DISEASE (FUMHD) WITH A FATAL COURSE

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A 7 year old boy was admitted with a sudden onset of painfull large ulceronecrotic lesions that developed out of pityriasis lichenoides (PL), accompanied by fever (38°). No other complaints.
Based on clinical grounds we made the diagnosis of FUMHD. However the biopsy showed infiltration of the dermis with large anaplastic CD30+ T-lymphocytes with loss of differention markers.
A staging before therapy did not show other locations, although this was not done very extensively due to the bad condition of the patient. Despite intensive care and cytostatic treatment his condition rapidly deteriorated and he finely died.
Discussion: CD30+ ALCL represents 10-15% of non Hodgkin lymphoma in children. Mostly it is a systemic disease, cutaneous forms usually present as an unique tumor and both forms have a good prognosis when treated (1).
The clinical picture of our patient was more that of FUMHD and before onset PL lesions were diagnosed. FUMHD is lethal in 25%, mostly by septic complications. Cases of FUMHD with CD30+ anaplastic T cells (2) have been described as well as clonality (3) and evolution to (3) or mimicking of (4) lymphoma mostly CD8+ T cell lymphoma.
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OP009 - SKIN LESIONS IN 100 POLISH PATIENTS WITH ATAXIA-TELANGIECTASIA

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Objectives: Ataxia-telengiectasia (AT) is a rare autosomal recessive disorder. It is classified as primary immunodeficiency associated with a defective DNA repair mechanism, but also belongs to phakomatosis.
AT is characterized by cerebellar ataxia, oculocutaneous teleangiectasias and immune deficiencies. Common skin lesions associated with AT include cutaneous telangiectasia, café-au-lait spots, skin atrophy and premature graying, seborrheic dermatitis, keratosis pilaris. Less frequent changes comprise: partial albinism and vitiligo, basal cell carcinomas, and atopic dermatitis.
Methods: In registry of Department of Immunology there are 1097 children with primary immunodeficiency disorders. Among them there are 100 AT patients: 47 females and 53 males with median age of 15,3 years.
Results: Oculocutaneous telangiectasia were observed in 98 and café-au-lait spots in 54 patients respectively. Progeric changes of hair and skin were ascertained in 76 patients. Seborrheic dermatitis is found in nearly all patients above 15 years. Vitiligo and partial albinism, a rare skin manifestations, were seen in 7 patients, atopic dermatitis in 3 and skin abscesses in 2 patients. No skin malignancies have been observed yet.
Conclusions: There is a variety of skin lesions in AT. The signs of premature ageing and oculocutaneous teleangiectasias are cardinal features of AT. They are supportive features in AT diagnosis. AT, as a DNA repair disorder, may constitute the model for further studies in mechanisms of skin cancer development.
References
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OP010 - GENERALIZED PEELING SKIN DISEASE: A MODEL DISORDER FOR ATOPIC DISEASES ?

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Generalized peeling skin disease with pruritus, and atopic diseases also referred to as peeling skin syndrome (PSS) type B [MIM 270300] is an unusual autosomal recessive ichthyosiform erythroderma characterized by lifelong patchy peeling of the entire skin.
It shares several clinical features with Netherton syndrome (NS). Moreover, both diseases appear very similar at the histological and ultrastructural level.
We studied a large consanguineous family with four affected individuals suffering from generalized peeling skin disease with severe pruritus and atopic manifestations, e. g. allergic reactions to food allergens such as fish or nuts. Hair shaft analysis did not reveal specific anomalies. Skin ultrastructure showed an enhanced detachment of corneocytes. To exclude the possibility of an allelic presentation of NS this diagnosis was excluded by mutational analysis of the entire SPINK5 gene that did not reveal any mutations.
We conclude that PSD is 1) genetically distinct from Netherton syndrome, 2) morphologically characterized by a profound defect of the epidermal barrier, and 3) may also represent a human model disorder for atopic diseases in general.

OP011 - CONGENITAL MELANOCYTIC NAEVI - A NEW SYNDROME

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Background: Congenital melanocytic naevi (CMNs) are known to be associated with neurological abnormalities, but have not been considered to be otherwise part of a developmental syndrome. In our paediatric pigmentary clinic it was noticed that many children with CMNs had a similar facial appearance, and a suggestion of different average growth pattern.
Methods: Facial photographs of 51 white European children seen in our clinic with CMNs of any site and size were selected from our image database on the basis of good full anterior facial photograph and neutral expression. They were reviewed independently by 2 clinical geneticists and scored using standardised categories for facial dysmorphology. Prevalences of individual features were compared statistically to known norms for a European population(1). Mean height, weight and head circumference data for 100 children with CMNs were analysed with respect to standard UK growth charts.
Results: CMNs are associated with characteristic facies, the most prevalent and significant features being a wide forehead, broad face, full cheeks, prominent everted lower lip, long or deep philtrum, short nose with broad nasal tip, periorbital fullness and eyebrow anomalies. The mean weight and height of boys with CMNs were greater than the population means.
Conclusions: We describe the new findings of characteristic facial features associated with CMNs, as well as different patterns of growth in this population. These findings help to define the Congenital Melanocytic Naevus syndrome, and may help to elucidate the aetiology of this condition.
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OP012 - HEREDITARY ANGIOEDEMA IN CHILDHOOD – PRESENTATION OF DATA FROM 15 DANISH CHILDREN

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Background: Hereditary angioedema (HAE) results from a deficiency of functional Complement C1 inhibitor and presents with different clinical signs including cutaneous angioedema and erythema marginatum. In childhood the disease can present with recurrent abdominal pain and there is an inherent risk of asphyxiation due to laryngeal edema. The prevalence is 1:71.000 and there is often a long diagnostic delay.
Objectives: To present data on Danish children with HAE.
Methods: A national Danish survey has been performed from 2001 to 2008 recruiting patients from hospital departments, dermatologists in private practice, Centres for Rare Diseases, the Danish patient organization and the national reference laboratory.
Results: 15 children among 82 Danish patients with HAE were identified.
Conclusion: The clinical manifestations, diagnostic difficulties, treatment options and family histories are presented together with some clues to the diagnosis. A diagnostic algorithm and management guideline for such patients will be proposed.
Reference
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OP013 - NO EVIDENCE FOUND THAT CHILDHOOD ONSET OF PSORIASIS INFLUENCES DISEASE SEVERITY, FUTURE BODY MASS INDEX OR TYPE OF TREATMENTS USED: A QUESTIONNAIRE AMONG 1926 PATIENTS

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Background. In more than one-third of the psoriatic population, the first manifestations occur in childhood. Whether the age of onset of psoriasis influences the march of psoriasis is not known.
Objectives. To describe the epidemiology and clinical features as well as prescribed treatments and familial distribution of childhood and adult onset psoriasis.
Methods. A structured questionnaire was sent to 5300 psoriatic patients. Respondents were divided in two groups: patients who experienced an onset of disease before the age of 18 (childhood onset psoriasis (COP)) and patients with an onset of disease from the age of 18 (adult onset psoriasis (AOP)).
Results. Questionnaires of 1926 (36.3%) patients were suitable for analysis. In 37.1% of patients first signs of the disease occurred before the age of 18. COP occurs predominantly in females, has a longer delay in diagnosis and a higher frequency of familial distribution. The development of guttate and erythrodermic psoriasis in adulthood is more frequently seen in COP. In contrast to the common believe, type of psoriasis in COP often remains the same from childhood to adulthood. There was no evidence found that getting psoriasis before the age of 18 years influences development of high BMI in adulthood, disease severity in later life or type of treatments used.
Conclusions. The age of onset of psoriasis does essentially not influence the subsequent course of the disease in adulthood.

OP014 - RICKETS IN CHILDREN WITH ICHTHYOSIFORM ERYTHRODERMA

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Background: Rickets is a disease of the growing bones due to various causes and vitamin D deficiency is the most common cause in India. Hyperparathyroidism and low vitamin D levels have been reported in few case reports of various ichthyoses and other keratinizing disorders. However there have been no large controlled studies to confirm this associa-

tion. Hence we undertook this study to find the occurrence of rickets in various ichthyosiform erythroderma.
Materials and Methods: Children with severe congenital ichthyoses and erythroderma due to keratinizing disorders were recruited. Age (±3 year), sex & season matched controls (Control group 1: children with skin diseases other than keratinizing didorders; Control group 2: Healthy volunteers) were included in the study. The study protocol included detailed history, examination, skeletal survey and baseline investigations including serum Ca/Po4, alkaline phosphatase , 25 (OH) vitamin D and parathyroid hormone
Results: Twenty nine cases (16 M, 13 F) were recruited which included EHK 10, NBIE 7, lamellar ichthyosis 5, ichthyosis vulgaris 4 , and psoriasis 3. Ten patients had clinically evident rickets and 12 of 26 had radiological features of rickets. Alkaline phosphatise was increased in 10 cases. The vitamin D levels were <9ng/ml in 16/25 (64%) patients and below 20 ng/ml in 24/25 (96%). PTH levels were >65pg/ml in 13/25 (52%) patients. (The results of the control group will be discussed at the time of presentation during the conference).
Conclusions: Our preliminary report suggests that children with severe congenital ichthyosis and ichthyosiform erythroderma are at increased risk of developing rickets. Such children should be screened and given vitamin D supplements to prevent deformities due to rickets.

OP015 - CHURG-STRAUSS SYNDROME IN CHILDHOOD

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Background: Churg-Strauss syndrome is a systemic necrotising eosinophilic vasculitis that is generally found in adults, but is extremely rare in children. We previously reported ten patients with Churg-Strauss syndrome but not in children.
Objectives: We investigated clinical findings and serological findings of childhood Churg-Strauss syndrome.
Methods: We described a 10-year-old Japanese girl who had Churg-Strauss syndrome without accompanying antineutrophil cytoplasmic antibodies and reviewed published cases in the English literature of Churg-Strauss syndrome in children.
Results: Twenty nine patients with Churg-Strauss syndrome in children were reviewed with an average patient age of 11.7 years, and an age range of 2 to 17 years. Our patient was not positive for antineutrophil cytoplasmic antibodies, and we detected only one patient with antineutrophil cytoplasmic antibodies in the 14 young patients with Churg-Strauss syndrome in our review. In contrast, more than half of adult patients with Churg-Strauss syndrome were positive for antineutrophil cytoplasmic antibodies. Our data showed a low incidence of nephritis in children with Churg-Strauss syndrome. Some reports have suggested that antineutrophil cytoplasmic antibodies-positive adult patients with Churg-Strauss syndrome are significantly correlated with nephritis compared to the antineutrophil cytoplasmic antibodies-negative patients.
Conclusions: Antineutrophil cytoplasmic antibodies seem to be the most significantly different between children and adults with Churg-Strauss syndrome. We propose that antineutrophil cytoplasmic antibodies might not be related to the pathogenesis of childhood Churg-Strauss syndrome, and suggest that the pediatric form of Churg-Strauss syndrome differs from the disorder in adults.

OP016 - MULTIPLE KERATOCYSTIC ODONTOGENIC TUMORS: MOSAIC PATTERN OF NEVOID BASAL CELL CARCINOMA SYNDROME

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Background: Keratocystic odontogenic tumors (KCOTs), formerly known as odontogenic keratocysts, are aggressive lesions of the jaw occurring either in isolation or in association with the nevoid basal cell carcinoma syndrome (NBCCS). Mutations in the PTCH1 gene are responsible for NBCCS and are also found in KTOCs, medulloblastomas and basocellular carcinomas in patients free from the syndrome (1). We describe a 13-year-old boy who presented multiple KCOTs. Complete examination and skull radiographs produced no evidence of any other NBCCS criteria. Neither of the parents had a history of basal cell carcinoma or any overt manifestation of NBCCS.
Objective: The aim of the present study was to to investigate the PTCH1 gene in the patient.
Methods: Genetic testing was performed on a KCOT, and skin and blood samples of the patient.
Results: We identified one PTCH1 frameshift mutation (c.2563_2570del8) in the KCOT sample. This frameshift mutation has been predicted to cause premature termination of PTCH1. The same mutation was also found in the blood samples but at a lower level than in the KCOT sample, suggesting a somatic mosaicism. The PTCH1 frameshift mutation, which was absent in the skin fibroblasts, exhibited a heterozygous pattern in the KCOT sample and in peripheral blood.
Conclusion: Molecular examination indicated that a novel PTCH1 mutation is involved in the pathogenesis of the patient's multiple KCOTs. The presence of the same mutation in KCOT and at lower levels in the blood samples demonstrated a mosaic pattern of NBCCS. To our knowledge this report is the second such case to be documented with a molecular study in the litterature(2).
References
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OP017 - ACUTE HEMORRHAGIC EDEMA OF INFANCY – A CASE SERIES OF 10 PATIENTS

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Background/Aims: Acute hemorrhagic edema of infancy (AHEI) is a relative unknown leukocytoclastic vasculitis. It consists of symmetric edema of the face and extremities with acute onset, followed by rapid development of ecchymotic purpura, with or without fever. Our objective was to determine the clinical features and outcome of AHEI and to discuss the complexity of the diagnosis.

Methods: Retrospective review of clinical files of patients diagnosed with AHEI in the last 9 years.
Results: We report 10 cases of AHEI (6 females; 4 males; median age at diagnosis of 11 months). Most cases occurred in the winter months. All had purpuric lesions in the inferior limbs and all except one had purpura elsewhere. Peripheral edema was only present in 7 patients and fever in 4; 7 had a context of previous infection, mostly upper respiratory tract, and 4 of those were still on active treatment with antibiotics/ antihistaminics. 3 patients complained of joint pain and one presented with blood in the stools, none had renal involvement. 8 patients had a diagnostic work-up to exclude other diagnosis such as sepsis but none had a positive urine or blood culture. Histological exam was performed in only 1 patient and was consistent with the clinical diagnosis. All had a full recovery in few weeks without specific treatment and only one had recurrent episodes.
Conclusion: Although typical, the clinical presentation of AHEI can be variable and sometimes be associated with life-threatening infections, raising issues of differential diagnosis. While the cutaneous lesions are certainly impressive, a wait-and-see approach is, in most cases, the only treatment required as complete resolution is expected in the following 2 to 3 weeks. AHEI is an under-diagnosed entity that is still unknown to many pediatricians. The obligation for skin biopsies and full septic screen should be reserved to the atypical cases.

OP018 - A RARE CASE OF CONGENITAL DEGOS DISEASE

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A 10-month old infant girl was referred to the Dermatology service for assessment of congenital skin lesions. She was generally unwell with progressive lethargy, irritability and failure to thrive, recurrent diarrhoea and unexplained abdominal distension. An acute abdomen led to laparotomy which showed oedema and adhesions within the mesenteric wall with no evidence of perforation or peritonitis. Bilateral ptosis was noted at the age of 4 weeks. Full body imaging showed evidence of brain calcifications, pleural effusions and ascites with mesenteric thickening. Laboratory investigations which included full blood count, haematinics, electrolytes, coagulation profile, auto-antibodies, viral serology and vasculitis screen only showed iron deficiency anaemia.
The skin lesions were multiple pink to red papules with central porcelain white atrophy and surrounding teleangiectasia distributed predominantly on the trunk suggestive of Degos disease. A skin biopsy showed features of medium vessel vasculopathy with no active vasculitis. Her condition continued to deteriorate and she died some months later. A post mortem examination showed widespread ulceration of both small and large bowel wall with multiple infarcts in the liver, spleen and brain with a vasculo-occlusive pathology consistent with Degos disease (malignant atrophic papulosis).
This patient had the systemic variant of Degos disease which may involve the skin, gut, brain, eyes and lungs. Our case demonstrates the importance of early recognition of this condition by its characteristic cutaneous lesions. To our knowledge, this is the youngest case of Degos disease reported in the literature.

OP019 - UNUSUAL DERMATITIS IN CDAGS SYNDROME: A DIFFERENTIAL DIAGNOSIS OF ACRODERMATITIS ENTEROPATHICA AND DYSMETABOLICA

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CDAGS syndrome is a rare autosomal recessive disorder delineated and mapped to chromosome 22q12-q13 in 2005. The acronym stands for craniosynostosis and clavicular hypoplasia, delayed closure of the fontanel, cranial defects, deafness, anal anomalies, genitourinary malformations, and skin eruption which, in some patients, has been classified as porokeratosis. The full spectrum of anomalies is rarely present.
A girl, first child of healthy non-consanguineous parents, was born at 34 weeks of gestation by caesarean section, following intrauterine cessation of growth. She presented with congenital craniosynostosis with turricephaly, anal atresia, and mild camptodactyly. Further course was complicated by failure to thrive and delay in motor development. At the age of 3 months, some weeks after weaning, well-demarcated, orange-red, crusted to scaly papules and plaques developed on her face, the extensor sides of the arms, and the perigenital area. In addition, pronounced diffuse alopecia was evident. Despite exclusion of zinc deficiency and other metabolic defects, oral zinc therapy was initiated but remained without effect. Histological examination revealed an unusual type of lichenoid dermatitis with abundant cytoid bodies and serous imbibition of the stratum corneum, not diagnostic for porokeratosis. The dermatitis ran an undulating course and was hardly responsive to various topical anti-inflammatory treatments.
In conclusion, a unique type of dermatitis resembling acrodermatitis enteropathica and dysmetabolica may serve as a diagnostic clue in CDAGS syndrome. Considering the 7 cases reported so far, histological signs of porokeratosis may be present or develop later.

OP020 - PAPULONECROTIC AND NODULAR TUBERCULID IN A IMMUNOCOMPROMISED CHILD: A CASE REPORT

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Tuberculous infection can mimic any other disease in an immunocompromised child and needs high index of suspicion. Here we report a child with tuberculous infection who presented as papulonecrotic and nodular tuberculid.
Our patient is a 5 year old Malay boy diagnosed Acute Lymphoblastic Leukaemia (ALL) in 2007 and currently on maintenance chemotherapy, presented with recurrent papules and nodules symmetrically on both upper and lower limbs for past 5 months. These lesions consisted of numerous crusted papules and a few reddish-skin coloured nodules. He also

had some pustules and papules on both feet. These lesions were itchy. He was empirically treated for scabies with no response.
A skin biopsy was performed and showed well formed caseating granulomas with inflammatory cell predominantly epitheliod cells and Langhans giant cells. Staining for acid-fast bacilli was negative. Tissue culture for Mycobacterium was negative. His Erythrocyte Sedimentation Rate was raised and his Mantoux was positive. His Chest X-ray showed pneumonic changes. He was started on four drugs regime of antituberculous therapy. His lesions responded favourably by 3 months and currently in complete resolution.
Tuberculous infection has various manifestations in immunocompromised children as reported in our patient and negative culture cannot exclude tuberculous infection.
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OP021 - (SIDE-) EFFECTS OF PROPRANOLOL FOR HEMANGIOMAS IN CHILDREN

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Abstract introduction: Infantile hemangiomas are frequently encountered tumors with a potentially complicated course. Recently propranolol has been described as a new and effective treatment option in hemangiomas.
Methods. Side-effects and special effects of propanolol are described in 23 children with complicated hemangiomas.
Results. All 23 patients with a (potentially) complicated course had a good response on propanolol treatment. Special effects: in 2 patients, steroid maintenance therapy could be tapered successfully after propranolol initiation. Furthermore in 3 older patients (12 and 15 months, 3 years old) propranolol proved to be an effective treatment option. Side-effects: 1 patient experienced a symptomatic hypoglycaemic event due to simultaneous use of propranolol and prednisone, 4 patients suffered from hypotension, 1 patient experienced bronchospasm after initiating propranolol and 4 patients suffered from restless sleep.
Conclusions. Also after the first year of life propranolol seems to be a good option for the treatment of (complicated) hemangiomas. Care should be taken for side-effects such as hypotension, bronchospasm and hypoglycaemia, especially with regard to patients on corticosteroid therapy, as these patients are at increased risk of hypoglycaemia. An advice for safe treatment of hemangiomas with propranolol is given.

OP022 - HODGKIN'S LYMPHOMA IN AUTOSOMAL RECESSIVE HYPERIMMUNOGLOBULIN E SYNDROME

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Background. Known hyper-IgE syndromes (HIES) include autosomal dominant (AD) HIES (Job's syndrome), caused by STAT3 deficiency, and autosomal recessive (AR) HIES, identified in 2004 and which genetic deficit is still unknown.
Case report. A 17-year-old white female was referred in consultation for poorly controlled, lifelong severe eczema that appeared in the first weeks of life. Past medical history included long-lasting asthma, hay fever, allergic gastroenteritis and recurrent sinus infections. Lymphocyte-predominant nodal Hodgkin's lymphoma had been diagnosed at 15, staged IIA and successfully treated with polychemotherapy and radiation therapy. Physical examination highlighted a well-nourished girl in no acute distress with widespread lichenified papules and plaques on the trunk and limbs. Routinary hematologic and immunology testing was unrevealing except for serum immunoglobulin (Ig) E levels of about 33000 IU/mL. The patient was prescribed topical corticosteroids, immunomodulators as well as antibiotics and antiseptics. On the basis of the National Institutes of Health scoring system, a diagnosis of HIES was achieved (score 24, cut-off of probable diagnosis: greater than 20). However, the patient lacked some common features of either STAT3-deficient, AD/sporadic HIES (e.g. characteristic facies, lung, musculoskeletal and dental abnormalities, and skin abscesses). As none of first- and second-degree relatives achieved a score equal or greater than 20, an AR trait was suspected.
Conclusion. While AD-HIES patients seems to be at higher risk of lymphoma, no cases of AR HIES-associated malignancies have been reported so far. We describe here the first case of AR HIES-associated malignancy and briefly review current knowledge on AR HIES.

OP023 - PROFILE OF CHILDHOOD VITILIGO IN KAYSERI/TURKEY: AN ANALYSIS OF 110 PATIENTS

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Vitiligo is a depigmentation disorder resulting from autoimmun destruction of cutaneus melanocytes. Half of the patients with vitiligo experience disease onset in childhood. Although largely similar to the disease in adults, pediatric vitiligo has differences in epidemiology, associations and treatment. The aim of this study to review the clinical characteristics of childhood vitiligo.Of the 110 children 50(45.5%) were boys and 60(54.5%) were girls.
The mean age of the patients was 9.6±3.8 years. The mean age of onset of the disease was 7.8±3.6 years. Generalized vitiligo was the most common type(70%), followed by localized(26.4%) and segmental types(3.6%). Fifteen patients(14.6%) had a family history of vitiligo. Ten(9.1%) had one family member, 5(%4.5) had more than one family member who was effected. The prevalence figures for vitiligo in first, second, third degree relatives of the probands were 10.9%, 1.8%, 0.9% respectively. Thyroid functions and thyroid autoantibodies were tested in only 71 patients. FreeT3 was decreased in 7 patients(9,9%), increased in 5 patients(%7), freeT4 was decreased in 5 patients(7%), thyroid stimulating hormone(TSH) was decreased in 1 patient(1,4%), increased in 17 patients(24,3%), antithyroglobulin was increased in 5 patients(5%), antiTPO was increa-

sed in 1 patient(1,5%). There was no accompanying disease in 88.2% of the cases. Hashimoto thyroiditis was observed in two patients. Asthma, atopic dermatitits, Down syndrome, hypothyroidi, epilepsy, autoimmune hemolitic anemia was detected in one patient each.We used local treatment in 64 patients(58,2%), narrowband ultraviolet B(nbUVB) in 21(19,1%), local+nbUVB in 19(17,3%), PUVA in 2(1,8%), local UVA in 1(0,9%), systemic treatment in 1 patient(0,9%). *Conclusion*: Vitiligo was higher in girls than boys and generalize vitiligo was the most common type of vitiligo.