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Irish Association of Dermatologists

Autumn Meeting

2nd & 3rd October, 2014

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Welcome Message from the President Dr Rosemarie Watson

I would like to extend a warm welcome to those of you attending our autumn meeting. I hope you enjoy our new venue in Athlone which should be a suitable location for dermatologists all over Ireland.

The theme for this meeting is hair. Hair- too much or too little- has been a great source of concern to mankind over the centuries. This has attracted the attention of both the beauty industry and health professionals sometimes producing treatments with very little evidence base. This meeting is very timely due to the recent major advances in this area. I am delighted to welcome our internationally renounced speakers and I am confident you will all acquire new and useful information for clinical practice. The satellite symposium on Acne and Rosacea will also I have no doubt add to our knowledge in this area.

We appreciate the support of our pharmaceutical sponsors without which we could not hold meetings of this calibre. I would also like to thank Jacqui Carroll and the executive committee and subcommittees who have been working very hard on your behalf this year. The plans are well advanced for our fiftieth anniversary celebrations next year so be sure to put April 23rd to 25th 2015 in your diary.

I hope you enjoy this meeting.

Dr Rosemarie Watson
President
IAD

Irish Association of Dermatologists
Autumn Meeting
2nd October 2014
Sheraton Hotel, Athlone

Galderma sponsored Symposium

Rosacea

10.30 – 10.40	Welcome and Introduction Professor Frank Powell
10.40 – 11.20	Pathophysiology of Erythema in Rosacea Professor Martin Steinhoff
11.20- 12.00	Management of the Patients Journey – current and future developments Dr. Laura Savage
12.00 – 12.30	Q & A

Galderma Biographical Sketches

Dr. Laura Savage, Leeds Dermatology Centre

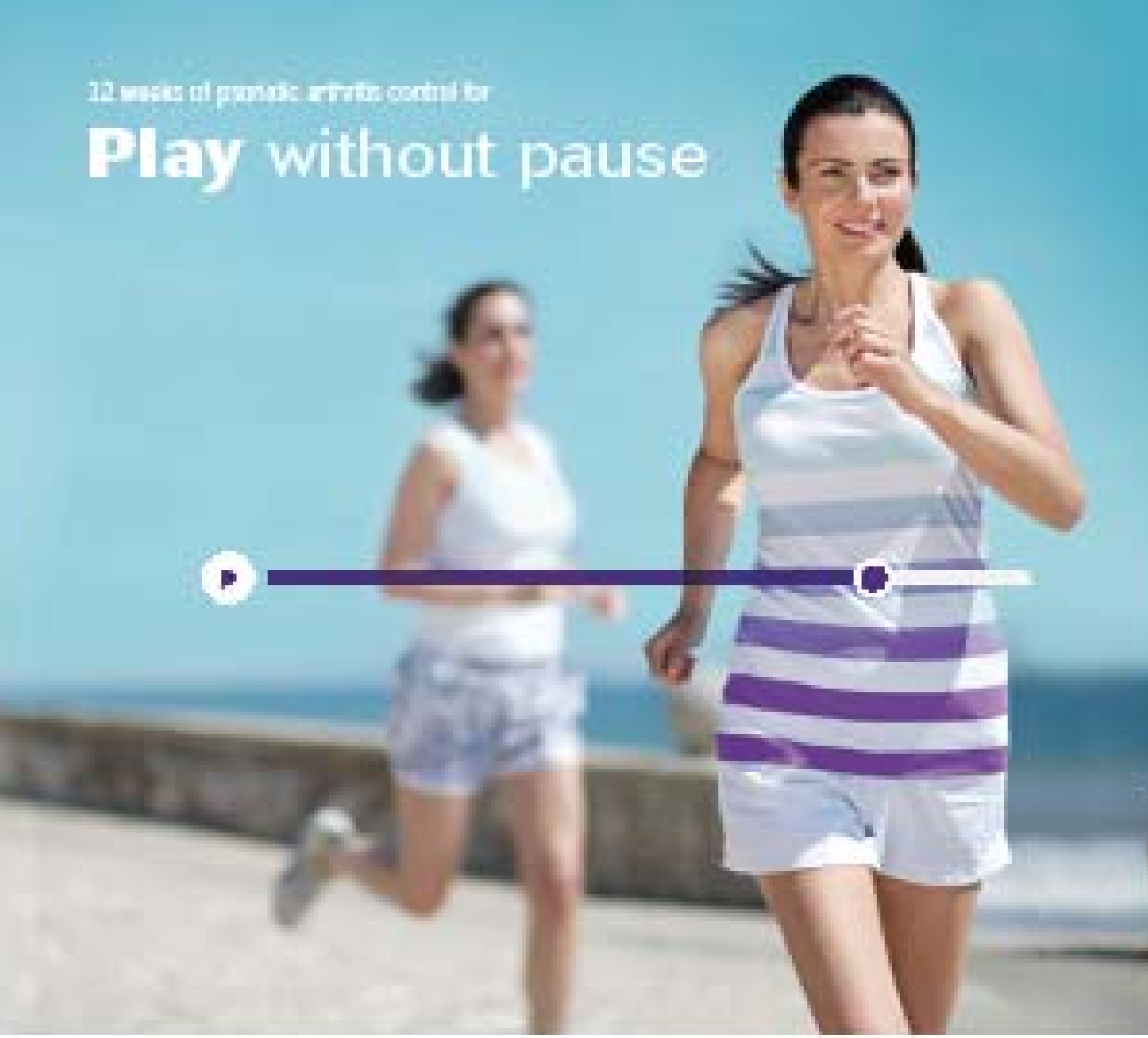
Dr Laura Savage graduated from the University of Edinburgh and trained in Dermatology within the Yorkshire Deanery. She was appointed as a Clinical Research Fellow in Dermato-Rheumatology in 2012 and is co-supervised by both Dermatologists (Dr Mark Goodfield, Dr Miriam Wittmann) and Rheumatologists (Professor Dennis McGonagle and Professor Paul Emery). Her research interests cross the boundaries of both specialties and relate to the development of strategies to detect early PsA in the dermatology arena in addition to the musculoskeletal response to skin-directed therapies. Her PhD specifically focuses upon the clinical and laboratory investigation/modulation of Th17 pathways in plaque psoriasis and subclinical psoriatic arthritis in both human tissue and mouse models. She is a member of GRAPPA and hopes to actively participate in collaborative research in the field of psoriatic disease following her PhD.

Prof Martin Steinhoff

Professor Steinhoff received his MD and MSc as well as PhD from the University of Marburg, Germany. In 2002, he became Assistant Professor of Dermatology at the University of Muenster, rising from assistant professor to full professor in only 6 years. He has board certifications in dermatology, venereology, phlebology and allergy from Germany, and in dermatology from the California Medical Board. Prof Steinhoff received several prestigious scientific awards for his research in Germany. To date, his group has published more than 200 articles, reviews and book chapters spanning basic science as well as clinical dermatology.

Prof Steinhoff is an established laboratory scientist who studies the substances and their receptors that cause inflammation, autoimmune disease or cancer of the skin. Beside general dermatology as a whole, his fields of clinical interests are eczema/itch, rosacea/acne, hives (urticaria) and wound healing (ulcers). He also performs and teaches sclerotherapy and foam sclerozation for the treatment or prevention of ulcers. He has run university consultative clinics for general dermatology, in-patient clinics, a day care unit and ulcer/phlebology clinic for these conditions in Germany for almost a decade.

Prof Steinhoff moved to the United States in January 2009 when he was appointed to the UCSF Departments of Dermatology and Surgery. Besides general dermatology, Prof Steinhoff established large clinics for itch/eczema as well as rosacea; two frequently occurring dermatological diseases with substantial impact on patient quality of life. He also established a successful NIH-funded research group for neuroimmunology. Prof Steinhoff is also principal investigator of various clinical trials, some of them based on his basic research results. Prof Steinhoff started his position as Professorial Chair of Dermatology and Director of the UCD Charles Institute at University College Dublin in January 2014.



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Irish Association of Dermatologists
Autumn meeting
Thursday 2nd & Friday 3rd October 2014

Thursday 2nd October 2014
Sheraton Hotel, Athlone

9.30am	Registration
10.30am-12.30pm	Galderma Satellite Symposium *"Rosacea"*
12.30pm-2.00pm	LUNCH & EXHIBITION

IAD PROGRAMME

2.00pm-2.45pm	Dr Andrew Messenger *'What's new in hair disease?'* Consultant Dermatologist Royal Hallemshire Hospital, Sheffield
2.45pm- 3.30pm	Dr Vicky Jolliffe *'Hair Loss Top Tips for Clinical Diagnosis'* Reader in Postgraduate Medical Education and Honorary Consultant Dermatologist Royal London Hospital/Queen Mary University of London, London.
3.30pm – 4.00pm	COFFEE & EXHIBITION
4.00pm –4.45pm	Dr Joe O'Connor & Dr Maurice Collins *'The Current Management of Androgenetic Alopecia'* Consultant Surgeons Hair Restoration Blackrock, Co. Dublin
5.00pm – 6.00pm	IAD Business Meeting
7.30pm	IAD CONFERENCE DINNER

Friday 3rd October 2014
Sheraton Hotel, Athlone

9.30 – 10.45am	Registrars' Symposium - Rogers Prize
10.45am	COFFEE & EXHIBITION
11.30 – 1.00pm	Case presentations
1.00pm – 2.00pm	LUNCH

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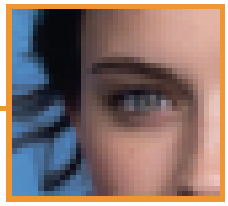

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Biographical Sketches

Dr Andrew Messenger
Andrew Messenger is a consultant dermatologist to Sheffield Teaching Hospitals and Sheffield Children's Hospital. He has a longstanding clinical and research interest in hair biology and disorders of hair growth. He was a founder member of the European Hair Research Society and its president 2004-6. He was president of the 7th World Congress of Hair Research held in Edinburgh in 2013.

Dr Joseph O'Connor MB, BCh, BAO, FRCS RCPS Glasg, FRCS Edin, FRCS Eng, FRCSI.
Consultant Surgeon
Dr O'Connor was educated at Rockwell College, studied Medicine at University College, Dublin and interned at St Vincent's Hospital. He was subsequently appointed lecturer at the University of Glasgow. He then spent a period as a researcher in cardiovascular and liver transplant surgery and has contributed to the international literature on these and other subjects. His Senior Registrar General Surgery rotation included a year as Vascular Fellow at the Cardiovascular Research Center, Seattle.

Dr. O'Connor holds a Specialist Certificate in General Surgery from the Royal College of Surgeons of England. He has over 30 years experience as a Consultant Surgeon and lecturer in surgery at postgraduate and undergraduate levels. He is a fellow of the Royal Colleges of Surgeons of Glasgow, Edinburgh, Ireland and England.

Dr Victoria M L Jolliffe MA (Cantab), FRCP, FRCS(Ed), MRCGP,ARCM
Honorary Consultant Dermatologist and Reader in Postgraduate Medical Education
Royal London Hospital/Queen Mary University of London. www.drvickyjolliffe.com
Vicky went up to King's College, Cambridge to read Classics before changing to Medical Sciences and completing her training at St Mary's Hospital London. Prior to completing CCST in Dermatology, Vicky worked in Accident and Emergency medicine and General Practice, providing her with a broad based medical background and understanding of the primary- secondary care interface.

An Honorary Consultant Dermatologist at the Royal London Hospital since 2004, she established a regional Hair Clinic in 2012. Educational Lead for the British Hair and Nail Society, she lectures internationally on Hair Disorders and is UK representative for the Pantene Hair Research Institute. She co-authored with Professor Rodney Sinclair 'Fast Facts-Disorders of the Hair and Nails' (2013). She has a special interest in e-learning and is UK Course Director for the Post Graduate Diploma in Clinical Dermatology, a yearlong web –based course aimed at General Practitioners(www.londondermatology.org). An accomplished String Player, her Alopecia UK fundraising video can be viewed on www.justgiving.com/quatuorVJ

Dr Maurice Collins MB, B.Ch, BAO, DLO, FRCSI, FRCS, FRCSEd.
Consultant Surgeon
Dr Collins is Medical Director and Team Principal of Hair Restoration Blackrock. He was educated at Belvedere College Dublin and did his undergraduate medical studies at University College Dublin. After graduating as a doctor he trained in General Surgery and received his Fellowship (FRCSI) in this specialty from the Royal College of Surgeons in Ireland. Dr Collins then undertook specialist surgical training in Ear, Nose and Throat Surgery and was awarded a Fellowship (FRCS) from the Royal College of Surgeons in London. He subsequently received a further Fellowship (FRCSEd.) in Head and Neck Surgery from the Royal College of Surgeons in Edinburgh. He has practised as a Consultant Surgeon in the Blackrock Clinic for the past 20 years.
Dr Collins' interest in hair transplant surgery started fifteen years ago and he has trained and studied internationally in this specialist subject with some of the best experts in the world. Dr. Collins and his team regularly attend, and participate in, the annual conferences of The International Society of Hair Restoration Surgery (ISHRS) and the European Society of Hair Restoration Surgery (ESHRS). Dr Collins contributes to the ISHRS Hair Transplant Forum International bi-monthly newsletter and was named Surgeon of the Month in May/June 2007.
Dr Collins has invited colleagues from around the world to HRBR in Dublin and has also attended numerous workshops in hair transplant surgery in both Europe and the United States.

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Irish Skin Foundation

Supporting people with skin conditions in Ireland

The Irish Skin Foundation strives to assist and support those affected with skin conditions via our community outreach activities. Below are just a few of our forthcoming events:

Allergy & Free From Expo:

Saturday 11th & Sunday 12th October, RDS Dublin *also*

Saturday 8th & Sunday 9th November, Cork City Hall

We will be on hand to support eczema sufferers, offering information, encouragement and assistance. 1-2-1 Dermatology Nurse advice available.

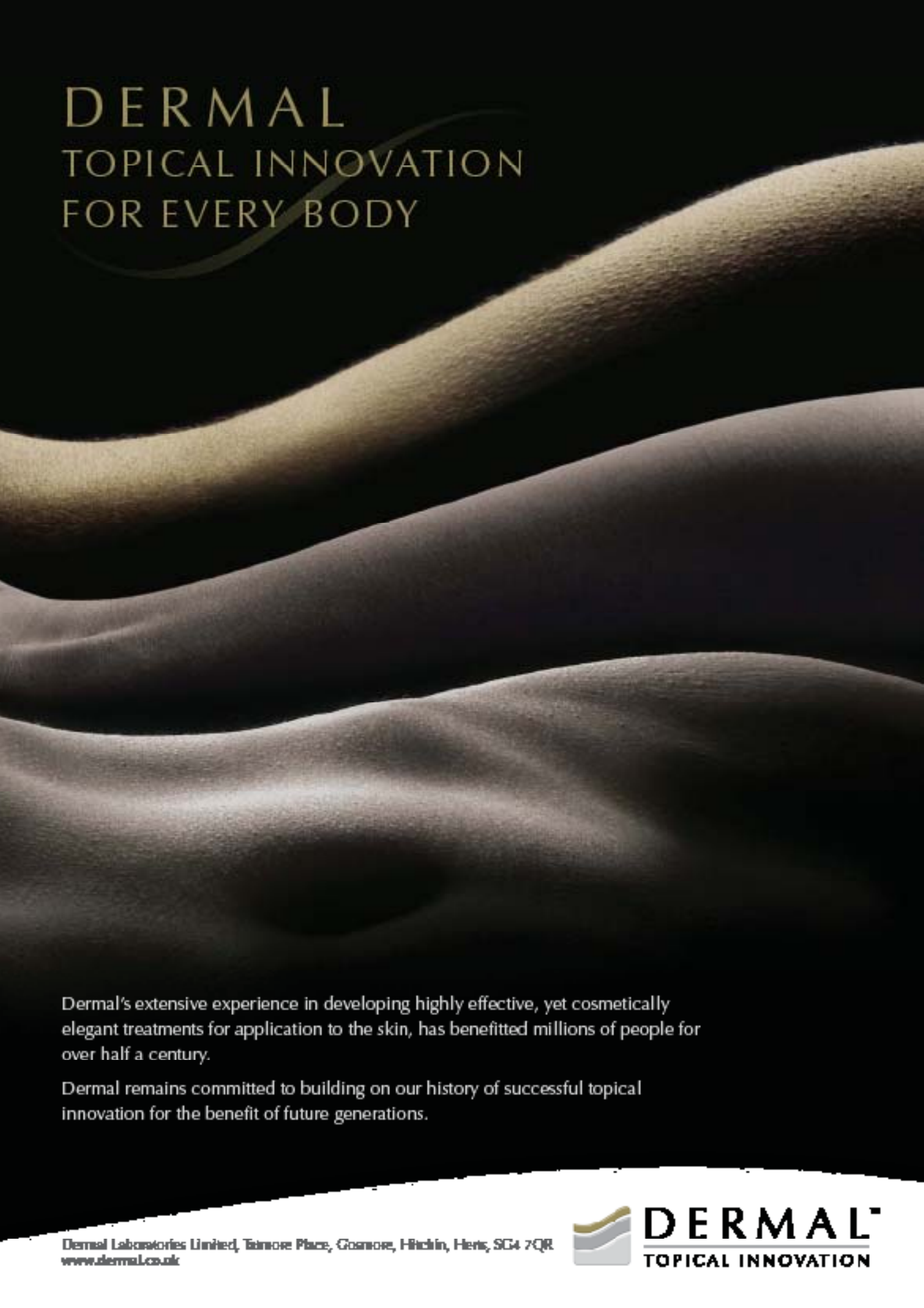
Cork Skin Awareness Event:

Saturday 15th November, The Kingsley Hotel, Victoria Cross, Cork

Free to attend public event where anyone affected by, or with an interest in skin can come along and learn more about conditions such as eczema, psoriasis, rosacea, skin cancer. A support forum where anyone with concerns about their skin can find help and advice.

Our mission is to support in all ways possible, to advocate on behalf of, to educate all involved with, and to bring comfort to those affected by skin disease in Ireland, their families and their carers.

Telephone: 01 716 6299 | www.irishskinfoundation.ie



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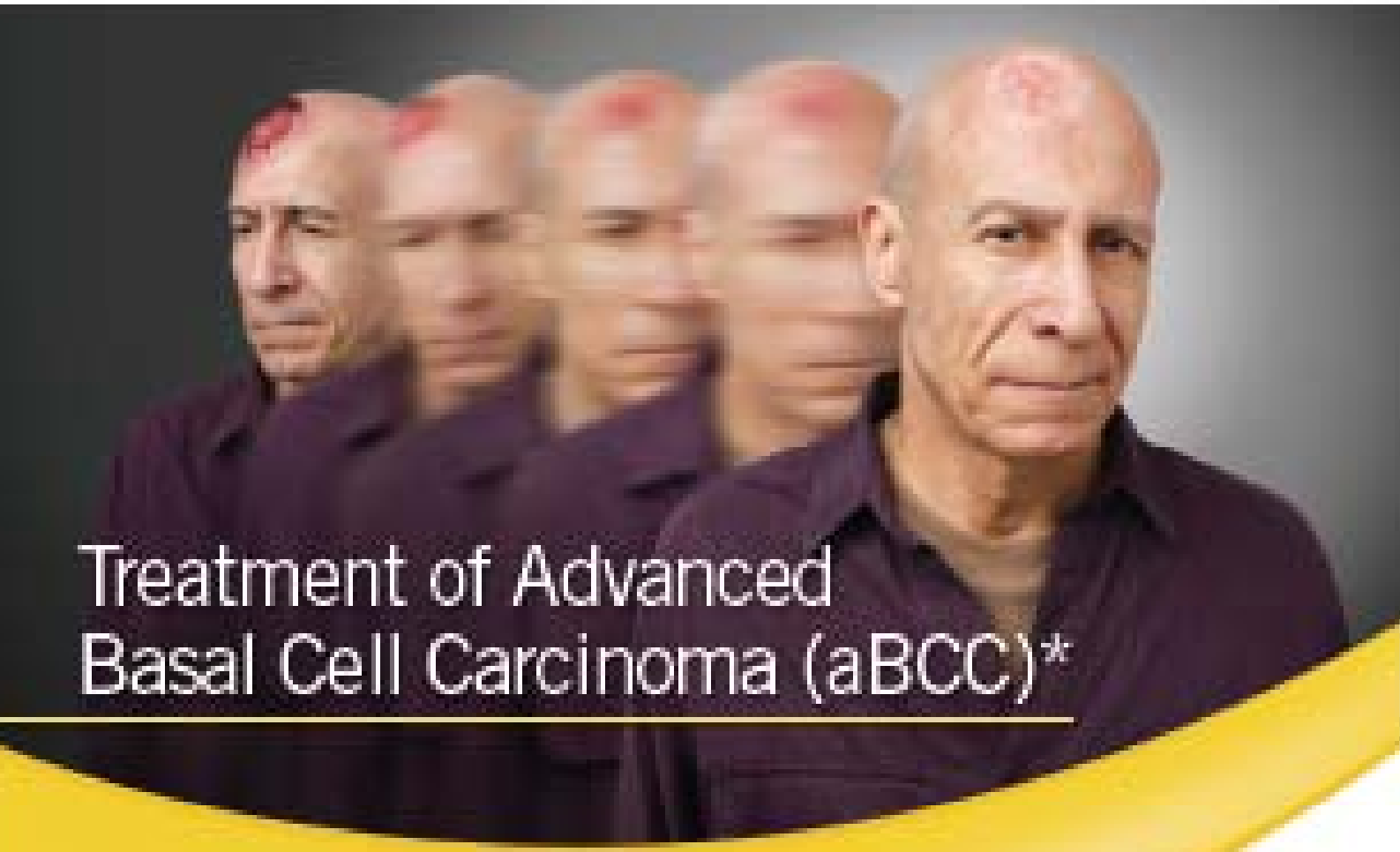

Treatment of Advanced Basal Cell Carcinoma (aBCC)*

Registrars' Symposium - Rogers Prize

01. 9.30am Assessment of a visual risk communication aid used to support patients in deciding about biological therapy.
M. Dolan, M. Connolly & A. Tobin

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02. 9.42am Attitudes Towards Sun Exposure In Inflammatory Bowel Disease Patients Taking Azathioprine.
E. Gilhooley, A. Farrelly, M Connolly, A. Tobin.

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03. 9.54am Patient perspectives on absolute and relative risk of cardiovascular disease in psoriasis using the QRISK algorithm.
R. Hellen, R. O'Connor, M. Connolly, AM. Tobin. Adelaide and Meath Hospital, Tallaght, Dublin 24

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04. 10.06am Hidradenitis Suppurativa and Crohn's Disease : A Case Series
S.Kirthi, R Hellen, R O'Connor, M Connolly, D McNamara, AM Tobin

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05. 10.18am Quality of life in Irish female patients with lichen sclerosus et atrophicus
NicDhonncha E, Foley CC, Laing M,Markham T, Murphy A, Marren P
Dermatology Department, University College Hospital, Galway

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06. 10.30am Clustered tender cheek nodules - a case of hereditary leiomyomatosis and renal cell cancer syndrome.
Clowry J, Collins P St Vincent's University Hospital, Elm Park, Dublin 4, Ireland

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Rogers Prize

■ Assessment of a visual risk communication aid used to support patients in deciding about biological therapy.

M. Dolan, M. Connolly & A. Tobin.

Introduction:
Communicating risk often proves challenging. Evidence indicates that people derive only the 'gist' from risk information, that assessment of risk can be influenced more by emotions than facts, and that people have a tendency to underestimate commonplace risk and overestimate rare risk. In this context, we decided to assess a visual risk communication aid (The Paling Perspective Scale ©) recently introduced to our practice as an adjunct to clinical consultation, to support patients in making choices with respect to two approved groups of biologic agents targeting tumour necrosis factor (TNF): anti-TNF monoclonal antibodies (adalimumab and infliximab), and sTNF receptors (etanercept).

Objectives:
We sought to investigate whether patients found this tool easy to understand, helpful in appreciating the potential side effects of treatment, and useful as an aid to decision making.

Methods:
The Paling Perspective Scale is a single page graphical representation, incorporating a histogram to portray risks of differing magnitude on a logarithmic scale. The risks associated with the TNF antagonists are depicted alongside more familiar risks such as death by motor vehicle in Ireland and risk of non-melanoma skin cancer, for comparative purposes to assist patients in evaluating the risks associated with treatment. The scale also incorporates verbal descriptors and includes a separate pictograph illustrating the risk of lymphoma expressed in a frequency format. Ten patients who received this scale as part of the decision making process were asked to complete a questionnaire.

Conclusions:
Ninety percent of patients found the scale easy to understand. All patients found the scale helpful in understanding the potential side effects of biological treatment. Seventy percent of patients thought the scale improved their ability to decide whether or not to commence a biologic treatment. However, patients documented the trust they still place in their doctor's view. The assessment of this tool is ongoing.

Evidence suggests that presenting probabilistic information graphically and numerically, promotes understanding and we propose ways the scale could be refined to further enhance patients' understanding of risk associated with biologics and inform their choices.

■ Attitudes Towards Sun Exposure In Inflammatory Bowel Disease Patients Taking Azathioprine.

E. Gilhooley, A. Farrelly, M Connolly, A. Tobin.

Introduction:
Immunosuppressive medications such as azathioprine are being used in patients with inflammatory bowel disease (IBD) patients in order to promote clinical remission[1]. Ongoing and past exposure to thiopurines, such as azathioprine, significantly increase the risk of non-melanoma skin cancer (NMSC) in patients with IBD, even before the age of 50 years[2].

Objective:
To examine the attitudes towards sun protection in patients taking azathioprine.

Methods:
Patients attending an inflammatory bowel disease outpatient clinic were asked to complete a questionnaire.

Results:
54% (27/50) of patients who completed this questionnaire were aware of the importance of sun protection measures with azathioprine. Of the 54% of patients aware of the need for sun protection measures only 18% (5) reported wearing sun protection factor on a daily basis. 46% (23/50) reported following sun protection strategies such as wearing protective clothing, hats and sunglasses. 52% (26/50) of patients who carried out this questionnaire frequently go on sun holidays. The Fitzpatrick skin classification of the patients sampled were as follows; Type I 22%, Type II 34%, Type III 32%, Type IV 10%, Type V 0%, Type VI 2%.

Conclusion:
IBD-specific, evidence-based guidelines for NMSC prevention have not yet been established. As for the general population, current recommendations include sun avoidance, use of broad-spectrum sun protection and minimisation of other risk factors of NMSC. Patients should be educated about the increased risk of NMSC at the initiation of immunosuppression and counselled on sun protection strategies.

References:
1. Pearson, D.C., et al., Azathioprine for maintaining remission of Crohn's disease. Cochrane Database Syst Rev, 2000(2): p. CD000067.

2. Peyrin-Biroulet, L., et al., Increased risk for nonmelanoma skin cancers in patients who receive thiopurines for inflammatory bowel disease. Gastroenterology, 2011. 141(5): p. 1621-28 e1-5.

■ Patient perspectives on absolute and relative risk of cardiovascular disease in psoriasis using the QRISK algorithm.

R. Hellen, R. O' Connor, M. Connolly, AM. Tobin.
The Adelaide and Meath Hospital, Tallaght, Dublin 24

Abstract:
Background: Psoriasis is an inflammatory condition with an increased risk of cardiovascular disease. The National Psoriasis Foundation recommends screening at least every 2 years for patients over 40. QRISK®2-2014 is a cardiovascular risk algorithm derived from a UK population1. It measures a patient's absolute risk of having a cardiovascular event over 10 years and their relative risk compared to healthy controls. It may perform as well as the Framingham algorithm and may have more accurate results as it considers ethnicity, body mass index, quantifies smoking and includes family history and co-morbidities. Rheumatoid arthritis is included as a

risk factor and we wished to see how this risk score would perform in patients with psoriasis. In addition, the equivalent "Heart Age" may be a beneficial tool for communicating relative risk to patients.

Methods:
18 patients have been recruited to date from a single dermatology department. We measured their height, weight, blood pressure, lipids and calculated PASI and DLQI. Absolute and relative risk of a cardiovascular event over 10 years was estimated using the QRISK®2-2014 score. Equivalent 'Heart Age' was also calculated. Patients completed questionnaires to determine their knowledge of co-morbidities, modifiable risk factors and the likely impact of relative risk on modifiable risk factors such as weight and smoking.

Results:
A total of 18 patients (66.6% men and 33.3% women) were included. Ages ranged from 30-62 years with an average of 46 years. No patients were classified as high risk. The average absolute 10-year risk of cardiovascular event was 6.88%. The average relative risk was 1.91. On average, patients scored 5.6 years older by their equivalent 'Heart Age'. 77.8% of patients were unaware that psoriasis is associated with greater cardiovascular mortality.

Conclusions:
QRISK®2-2014 is a beneficial tool in estimating cardiovascular risk in psoriasis patients and identifies modifiable risk factors essential for the primary prevention of cardiovascular events. Many of our patients were not aware that psoriasis is associated with increased cardiovascular mortality. Relative risk presented as an equivalent 'Heart Age' may be a beneficial adjunct to motivate changes in lifestyle and aid clinicians in screening for cardiovascular risk factors.

1: Hippisley-Cox J et al. HYPERLINK "http://www.ncbi.nlm.nih.gov/pubmed/17615182" Derivation and validation of QRISK, a new cardiovascular disease risk score for the United Kingdom: prospective open cohort study. BMJ. 2007 Jul 21;335(7611):136.

■ Hidradenitis Suppurativa and Crohn's Disease : A Case Series

S.Kirthi, R Hellen, R O'Connor, M Connolly, D McNamara, AM Tobin

Introduction:
Hidradenitis Suppurativa (HS) is a chronic inflammatory skin condition characterized by recurrent, painful abscesses, nodules and draining sinus tracts with bands of severe scar formation. Cutaneous Crohn's Disease (CD) may also present with similar skin lesions and CD and HS occur together at a rate that varies from 0.6% to 38% based on isolated case reports. A recent cytokine and leukocyte profiling by H.H van der Zee et al demonstrated raised TNFα, IL-β and IL-10 in tissue samples of HS patients suggesting a common underlying pathology for both conditions1.

Aims:
We wished to examine the overlapping syndrome of Crohn's Disease and Hidradenitis Suppurativa in an Irish cohort.

Methods:
Cases with HS and CD were identified by HIPE Code at Tallaght Hospital from 1990-2014. A retrospective chart review was performed of all cases.

Results:
In total, 4 patients with both HS and CD were identified. 50% were female. The median age of diagnosis for both conditions was 31 years. In all 4 cases, CD had preceded the diagnosis of HS, with a median interval of 34.5 months to HS diagnosis. 100% of patients smoked. Of note, 50% of patients had additional autoimmune conditions, 1; psoriasis and pyoderma gangrenosum, 1; ankylosing spondylitis. Despite a high BMI being associated with HS, only 1 patient (25%) in this cohort had a BMI of ›30. Of note, no patients had a family history of HS. All patients required treatment with a TNF-alpha inhibitor in addition to standard antimicrobial therapy. 75% of patients (3 of 4) had an improvement of Hurley's score on commencing anti-TNF therapy. This is the largest case series to date reported in the literature for an Irish cohort to our knowledge.

Conclusion:
Our cohort suggests that combined HS and CD syndrome affects young smokers and is frequently associated with other autoimmune conditions. Most will require anti-TNF alpha therapy to control symptoms. For those who do not respond, new therapeutic agents are eagerly sought, and further investigation with regard to interleukin 1 blockade is definitely worth investigating to treat combined CD and HS.

Reference:
1. 1.Br J Dermatol. 2011 Jun;164(6):1292-8. Elevated levels of tumour necrosis factor (TNF)-α, interleukin (IL)-1β and IL-10 in hidradenitis suppurativa skin:a rationale for targeting TNF-α and IL-1β. van der Zee HH1, de Ruiter L, van den Broecke DG, Dik WA, Laman JD,

■ Quality of life in Irish female patients with lichen sclerosus et atrophicus

NicDhonncha E, Foley CC, Laing M,Markham T, Murphy A, Marren P
Dermatology Department, University College Hospital, Galway

Introduction:
Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory dermatosis, more commonly occurring in women, which mainly affects the anogenital region. The most frequently reported symptoms are pruritus, discomfort, dysuria and dyspareunia. To date, there is little published data on the effect of LSA on patient quality of life (QOL).

Objectives:
To investigate the health-related QOL of a sample of Irish women with LSA using standard screening questionnaires.

Methods:
Patients attending our hospital with a diagnosis of LSA were identified from the hospital letter database (n=102). Patients were excluded if they had never attended Dermatology (n=4), if they had no pending Dermatology appointment (n=54) or if they did not have biopsy-proven LSA (n=8). The remaining patients (n=36) were offered the opportunity to participate in the study between January 2014 and June 2014. The included patients completed 2 anonymous questionnaires – the Dermatology Quality of Life Index (DLQI) and the Skindex-29.

Results:
Complete data was available on 26 patients with biopsy-proven LSA. Median age at presentation was 59.2 years (mean 55.2, range

12.5-76.8). Median duration of symptoms at time of initial presentation was 2 years (mean 3.6, range 0.3-10).
DLQI scores ranged between 0-10, with a mean score of 3.31. The impairment in QOL was none in 9/26 patients (35%), mild in 13/26 (50%) and moderate in 4/26 patients (15%). No patients reported severe impairment in QOL.
The studied patients had a mean total Skindex-29 score of 26.57 indicating mild impairment of health-related QOL overall. Severe impairment (score ›44) was reported in 4/26 (15%), moderate (score 32-43) in 6/26 (23%), mild (score 25-31) in 4/26 (15%) and little (score‹25) in 12/26 (46%). Domain scores for symptoms, emotions, and functioning were 37.78, 24.42, and 21.23 respectively.

Conclusion:
Lichen sclerosus et atrophicus is associated with impairment of quality of life in our patient population. While the majority of patients had mild impairment in quality of life, a smaller proportion of patients had severe impairment in quality of life. Previously published studies have shown at least moderate impairment in quality of life in association with LSA1.

References:
1. Lansdorp CA, van den Hondel KE, Korfage IJ, van Gestel MJ, van der Meijden WI. Quality of life in Dutch women with lichen sclerosus. Br J Dermatol 2013 Apr;168(4):787-93

■ Clustered tender cheek nodules – a case of hereditary leiomyomatosis and renal cell cancer syndrome.

Clowry J, Collins P
St Vincent's University Hospital, Elm Park, Dublin 4, Ireland

Introduction:
A 41 year old male presented with an approximate 20 year history of multiple, non mobile, firm, flesh coloured folliculocentric nodules in a clustered distribution on his lateral right cheek. The size of the lesions ranged from from 3-5mm. These were occasionally tender on exposure to the cold. He had no similar lesions elsewhere and was otherwise asymptomatic. He had no additional medical history. Family history was notable for early onset uterine fibroids in the patient's sister and a paternal aunt, who also developed uterine and renal cancer in her 70s. He had two children who were healthy and well.

Histopathology revealed a benign dermal spindle cell lesion, consisting of bundles of smooth muscle cells with cigar-shaped nuclei. The appearances were consistent with a benign piloleiomyoma.

The patient proceeded to have sequence analysis of genomic DNA. Results demonstrated heterozygosity for the c.301C›T; p(Arg101*) pathogenic mutation in exon 3 of the fumarate hydratase gene. This is an autosomal dominant mutation consistent with a diagnosis of Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) syndrome.

This is a rare autosomal dominant condition characterised by cutaneous and uterine leiomyomata and renal tumours in 10-16% of cases.1
Renal cell cancer in HLRCC presents as a solitary, unilateral lesion but may be aggressive and metastatic at diagnosis.

Patients often first present to dermatologists, since cutaneous leiomyomata are the earliest and most common manifestation. The mean age of onset is 25 years.2 They are found in approximately 76% of cases,2 although a higher penetrance has been reported in males.1

To date, the patient has been referred for genetic counselling and has had a normal renal ultrasound. He has declined surgical excision of his cutaneous piloleiomyomata. Ongoing surveillance has been arranged with the urology service.

1. Alam NA, Barclay E, Rowan AJ, Tyrer JP, Calonje E, Manek S, et al. Clinical features of multiple cutaneous and uterine leiomyomatosis: an underdiagnosed tumor syndrome. Arch Dermatol. 2005;141(2):199-206

2. Pithukpakorn M, Toro JR. Hereditary Leiomyomatosis and Renal Cell Cancer. In: Pagon RA, Adam MP, Ardinger HH, Bird TD, Dolan CR, Fong CT, et al., editors. GeneReviews(R). Seattle (WA): University of Washington, Seattle

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Laboratoires dermatologiques Avène PARIS

Pierre Fabre

Case presentations

01. 11.30am
Congenital segmental alopecia - Atypical Focal Facial Dermal Hypoplasia?
Dr Lorraine Jennings1, Dr Alan Irvine2 and Dr Sinead Collins1
1Department of Dermatology, Our Lady of Lourdes Hospital, Drogheda, Co. Louth
2 Department of Dermatology, Our Ladies Children's Hospital, Crumlin, Dublin

02. 11.40am
Hirsutism in a teenager
MA McAleer, E O' Dea, R Watson Our Lady's Children's Hospital Crumlin

03. 11.50am
Takotsubo cardiomyopathy and telogen effluvium associated with severe broncho-pneumonia
S. McCarthy, I. McDonald, C. Fahy, N. Mahon, F.J. Moloney
Department of Dermatology ,Mater Misericordiae University Hospital

04. 12.00pm
Successful treatment of a patient with Chronic Mucocutaneous Candidiasis due to a gain-of-function mutation in STAT-1 with JAK1/2 inhibitor ruxolitinib.
Eleanor Higgins1 Conleth Feighery2, Maeve McAleer3 Desa Lilic 4 AlanD.Irvine1,3, 5.

[1] Dermatology [2] Immunology, St James's Hospital,
[3] National Children's Research Centre, Our Lady's Children's Hospital, Dublin [4] Primary Immunodeficiency Group, Institute of Cellular Medicine Newcastle University, UK.
[5] Department of Clinical Medicine, Trinity College Dublin.

05. 12.10pm
Lichen Planopilaris with Facial Papules
A. Flynn, B. Wynne. St James's Hospital, Dublin

06. 12.20pm
Kerion induced scarring Alopecia
Flynn, B. Wynne. St James's Hospital, Dublin

07. 12.30pm
A newborn referred with blistering
W. Abdelrahman, S Clements, S Hoey
Dept of Dermatology, Royal Victoria Hospital, Belfast, Northern Ireland

08. 12.40pm
Treatment of cutaneous metastatic melanoma with topical diphencyprone - a case report
I.McDonald, F.M. Keane, J.A. McCaffrey, F.J. Moloney
Department of Dermatology, Mater Misericordiae University Hospital

09. 12.50pm
A case of melanonychia in a child.
RH El-khayat1, M Y Walsh2, A Rashid3, K McKenna1 .
[1.] Department of Dermatology [2.] Department of Pathology
[3.] Department of Plastic Surgery. Belfast Health and Social Care Trust.

Registrars Symposium Case presentations

Congenital segmental alopecia - Atypical Focal Facial Dermal Hypoplasia?

Dr Lorraine Jennings[1], Dr Alan Irvine[2] and Dr Sinead Collins[1]
Department of Dermatology, Our Lady of Lourdes Hospital, Drogheda, Co. Louth
Department of Dermatology, Our Ladies Children's Hospital, Crumlin, Dublin

Abstract:
A 13 year-old South-African male presented to our Paediatric Dermatology clinic with right-sided scalp alopecia that had been present since birth. He was the product of spontaneous normal delivery. There was no relevant past medical or family history. His parents were non-consanguineous.

Examination revealed a well-demarcated whorled pattern of alopecia on the right parietal scalp and vertex, associated with an asymmetrical underlying bony prominence. There were several circular atrophic scars with anetoderma in a linear pattern along the edge of the parietal alopecia, which had also been present since birth. An area of alopecia was noted extending to the right eyebrow. All sites of alopecia-involved skin had a doughy, extensile consistency. The patient was otherwise developmentally normal. His nails and teeth were normal, except for some bilateral incisor overcrowding. No skeletal abnormalities were detected.

Diagnostic punch biopsies were performed from the affected scalp and from an area of clinically unaffected skin. There were no visible hair follicles in the affected areas, and no apparent difference in elastic staining between the samples

Congenital alopecia is associated with a wide range of genetic conditions or may be a marker for an underlying metabolic disorder, which may impact on the mental and physical development of a child, empasising the importance of early diagnosis.

This case represents a congenital, unilateral, scarring alopecia with underlying soft tissue abnormality, in an otherwise healthy teenager. The distribution and atrophic scarring is suggestive of focal facial dermal hypoplasia (FFDH) Type III (Setlies Syndrome),1 a rare form of ectodermal dysplasia. However, the associated textural changes seen in this case are atypical and not previously reported in this condition. A single case report of localised Ehlers-Danlos2 exists also, with similar features to our case and without any systemic features of the disease.

We present this child with segmental alopecia with hyperextensibilty as an atypical case of focal dermal hypoplasia in the absence of a more unifying diagnosis. Further diagnostic suggestions are welcome.

References:
Setleis H, Kramer B, Valcarcel M, Einhorn AH. Congenital ectodermal dysplasia of the face. Pediatrics 1963;32:540–8.
Localized Ehlers-Danlos syndrome. HYPERLINK "http://www.ncbi.nlm.nih.gov/pubmed?term=Cullen%20SI%5BAuthor%5D&cauthor=true&cauthor_uid=434850"Cullen SI. Arch Dermatol. 1979 Mar;115(3):332-3.

Hirsutism in a teenager

MA McAleer, E O' Dea, R Watson, Our Lady's Children's Hospital Crumlin

Abstract:
A 15 year-old girl presented with severe hirsutism. She had insulin resistance secondary to a mutation in the tyrosine kinase domain of the insulin receptor. She was obese and had polycystic ovarian syndrome.

The patient had previously tried to improve her hirsutism with intense pulsed light treatment at a local laser centre without success. Anti-androgen therapy had been ineffective. Epilation in conjunction with topical eflornithine 15% cream had also proved unsuccessful.

The teenager suffered severe psychological morbidity and social difficulties secondary to the hirsutism. She reported anxiety and depressive symptoms. When she presented to our service she had ben avoiding leaving her bedroom for a year, which had a considerable impact on her schooling.

Treatment was commenced with the Alexandrite 775nm laser to the most visibly evident hirsute areas of the face, neck and upper chest. The patient has had 9 treatments to date. There has been an excellent improvement, with an 80% reduction in hair. The patient's psychological health has significantly improved. She has returned to school and can socialise with her peers.

Multiple studies have demonstrated effective hair removal with the long-pulsed alexandrite laser at fluences of 10-40 j/cm2 and pulse durations of 2-20 ms. At fluences of 20-40 j/cm2, several studies have reported hair reduction of 70-80% after multiple (at least 3-5) treatments.

In adolescent females hirsutism is associated with a decreased quality of life, a higher prevalence of anxiety disorder, and lower self-esteem.1 Higher levels of hair growth have been significantly correlated with a lower quality of life and symptoms of anxiety and depression. Laser treatment of facial hair in 70 women significantly improved their quality of life scores.[2]

Our case highlights the importance of early and effective treatment of hirsutism, particularly in adolescents, in an attempt to reduce the adverse psychological and social impact of the disease.

1.Drosdzol A Skrzypulec V, Plinta R. Quality of life, mental health and self –esteem in hirsuite adolescent females. JPOG. 2010;31:168-175
2.Maziar A et al. Unwanted facial hair removal with laser improves quality of life in of patients. J Cosmet Laser Ther 2010;12:7-9

Takotsubo cardiomyopathy and telogen effluvium associated with severe broncho-pneumonia

S. McCarthy, I. McDonald, C. Fahy, N. Mahon, F.J. Moloney
Department of Dermatology,Mater Misericordiae University Hospital

Introduction:
The pathophysiology of takotsubo cardiomyopathy (broken-heart syndrome) is poorly understood. Both it and telogen effluvium are reactive processes preceded by similar triggers including emotional and physical stressors, or medications. We describe the first report of both conditions occurring concurrently following an episode of severe bronchopneumonia.

Case History:
A 67-year-old female was admitted to a tertiary referral centre in March 2013 with community-acquired pneumonia secondary to haemophilus influenza. She was treated with vancomycin, meropenem and clarithromycin and was discharged six days later. She re-presented two weeks later with symptomatic persistent lingular pneumonia and was treated with moxifloxacin. Six weeks post discharge, the patient presented with sudden onset of dyspnoea. ST elevation was noted on her ECG antero-laterally. Coronary angiogram showed extensive antero-apical hypokinesia, with little coronary artery disease, consistent with takotsubo cardiomyopathy. During her inpatient stay she noted sudden onset diffuse scalp hair shedding. She had no personal or family history of alopecia. Examination revealed hair thinning over the vertex and frontal scalp with no inflammation or scarring evident. She had a positive hair pull test with increased numbers of telogen hairs. Thyroid function and iron stores were normal. A clinical diagnosis of acute telogen effluvium was made and the patient reassured. At clinic review four months post discharge she had full hair regrowth. By that time, her cardiac function had returned to normal.

Discussion:
Medications have been implicated in cases of both both telogen effluvium and takotsubo cardiomyopathy, however, the presumptive trigger in our case was her severe bronchopneumonia and resultant stresses. While the interval between inciting event and onset of hair shedding is dictated by the individual patient's telogen hair cycle length, the time scale in relation to development of takotsubo cardiomyopathy, after exposure to an emotional or physical stress, is less predictable. Both resolved without sequelae, an

Registrars Symposium Case presentations

indication of the favourable prognosis seen with both diagnoses. The two conditions developing in concurrence and the temporal association with her illness suggest that the same stress-induced catecholamine release, with toxicity to and subsequent stunning of the myocardium thought to result in broken-heart syndrome may involve similar neurotransmitter and hormonal pathway changes inducing catagen in the hair follicle.[1]

1. Satoshi Kurisu, Yasuki Kihara, Tako-tsubo cardiomyopathy: Clinical presentation and underlying mechanism, Journal of Cardiology, Volume 60 Issue 6, December 2012, Pages 429-437.

Successful treatment of a patient with Chronic Mucocutaneous Candidiasis due to a gain-of-function mutation in STAT-1 with JAK1/2 inhibitor ruxolitinib.

Eleanor Higgins1 Conleth Feighery[2], Maeve McAleer[3] Desa Lilic [4] AlanD. Irvine[1,3, 5.]
[1]Dermatology [2]Immunology, St James's Hospital, [3]National Children's Research Centre, Our Lady's Children's Hospital, Dublin
[4]Primary Immunodeficiency Group, Institute of Cellular Medicine Newcastle University, UK. [5]Department of Clinical Medicine, Trinity College Dublin.

Abstract:
RW.CITE{{194 Xing,L. 2014}} (1) demonstrated the role of cytotoxic T lymphocytes in alopecia areata (AA) and provided important mechanistic information on the pathogenic T-cell inflammatory pathways in this autoimmune condition. They described three patients with AA successfully treated with the oral Janus kinase (JAK) family protein tyrosine kinase inhibitor ruxolitinib. We further show the potential utility of ruxolitinib for AA occurring in the wider context of a genetic autoimmune/immunodeficiency syndrome. We recently treated AA associated with chronic mucocutaneous candidiasis (OMIM #614162) in a 28-year-old female with ruxolitinib. Our patient had a sporadic autosomal dominant gain-of-function (GOF) mutation (heterozygous c1159A>G (p.(Thr387Ala) in exon 14 of STAT1 and had a two-year history of alopecia areata. This had progressed despite previous intralesional corticosteroid injections, with greater than 40% scalp involvement. She also had recalcitrant oral candidiasis since childhood, with frequent flares despite daily oral fluconazole.

STAT1 gain-of-function with such mutations may be mediated via enhanced phosphorylation resulting in gain of function of gamma interferon activating factor and subsequent aberrant IL-17 immunity. Patients with gain-of-function STAT1 mutations also have decreased STAT3 that may explain downstream reduction of IL-17 expression. In canonical IFN-γ-JAK-STAT1 signalling, ligand engagement of the IFN-γ receptor leads to activation of receptor-associated JAK1 and JAK2.

We therefore hypothesized that JAK1/2 inhibition would target this pathway and would ameliorate both the CMC and the associated autoimmune AA phenotype in our patient. Ruxolitinib is a JAK 1/2 inhibitor approved for treatment of myelofibrosis, targeting pathogenic mutant JAK 2 pathway signalling and activation of transcription (JAK-STAT) pathways. We treated our patient with ruxolitinib 20mb twice daily for twelve weeks (off-licence). After 2 weeks, our patient experienced dramatic hair regrowth in all affected areas of alopecia. The hair regrowth was uniform and thick, with full regrowth at completion of treatment and this was sustained on review 3 months after completion of therapy. Our patient also reported complete resolution of oral candidiasis while on ruxolitinib treatment.

We hypothesize that correction of the GOF-STAT1 may allow restoration of STAT3-dependent production of IL-17, thus improving oral candidiasis. Xing et al highlight the potential for further clinical evaluation for ruxolitinib and other JAK inhibitors in the treatment of AA. Our case suggests the potential therapeutic benefit of ruxolitinib in GOF-STAT1 genetic immunodeficiency/autoimmunity syndrome. While further evaluation on additional patients in clearly needed, this therapy may represent an ideal personalized treatment for patients with gain-of-function mutations in STAT1.

1. Xing L, Dai Z, Jabbari A, Cerise JE, Higgins CA, Gong W, et al. Alopecia areata is driven by cytotoxic T lymphocytes and is reversed by JAK inhibition. Nat Med. 2014 Aug 17.

LICHEN PLANOPILARIS WITH FACIAL PAPULES

Dr Aoibheann Flyee, Dr Bairbre Wynne
St. James's Hospital, Dublin

Case Summary:
Lichen planopilaris (LPP) is a rare inflammatory condition that results in patchy progressive permanent hair loss mainly on the scalp. It commonly affects young adult females. Frontal Fibrosing Alopecia is considered a particular clinical form of LPP that primarily involves the scalp hair over the frontal hairline. Concomitant papules at different body sites are a well-recognised feature. However, facial papules associated with Lichen Planopilaris have only been described once in the literature. We a case of Lichen Planopilaris with facial papules.

MH, a 44-year-old woman presented with a 1-year history of facial and scalp irritation. Her General Practitioner had treated her with protopic, eumovate and diprosalic for presumed psoriais. Clinical examination revealed scarring alopecia along her frontal scalp with perifollicular erythema. There was minimal scale throughout the rest of her scalp. She also was noted to have loss of the lateral third of her eyebrows, along with prominent papules on her temples. Histopathology was consistent with lichen planopilaris. She is managed on Hydroxycholoroquine orally and Isotretinoin topically to the papules.

References:
Facial papules in Frontal Fibrosing Alopecia: Evidence of Vellus Follice Involvement.
Donati A, Molina L, Doche I, Valente NS, Romiti R
Arch Dermatology 2011 Dec; 147(12): 1414-7

Kerion induced scarring Alopecia

Flynn, B. Wynne. St James's Hospital, Dublin

Case Summary:
We present a case of a 2-year-old boy with significant scarring alopecia due to a large kerion on his scalp.

JS age 2 years 4 months presented to Accident and Emergency with a 7-month history of a progressive scalp kerion. He had been seen by a doctor not on the specialist registrar for dermatology. He treated him with topical steroids and it continued to progress until his presentation to hospital. The week prior to admission he had been unable to sleep due to the pain on his scalp, which was now oozing and bleeding. Clinical examination revealed a large Kerion with satellite lesions and cervical lymphadenopathy. Fungal scrapings grew Microsporium Canis. He was commenced on Oral Itraconazole, Nizoral Shampoo, Oral Flucloxacillin, Oral Prednisolone and Paraffin Gel. Within two weeks his scalp had significantly improved, with some mild swelling and minimal crusting. He unfortunately has a large area of scarring alopecia on his parietal scalp. He has been continued on the same treatment with the addition of topical fucidin.

This case highlights the importance of Dermatology review for persistent tinea capitis/kerion. Due to the delay in appropriate management this child has a large area of scarring alopecia on his parietal scalp. Given the cosmetic and psychological effects this will have long-term, he may be a suitable candidate for hair transplantation.

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Permanent Hair Loss after Kerion Celsi
Bonven TF, Iversen E, Kragballe K
Ugeskr Laeger 1991 Nov 4;153(45):3151-2

Hair Loss Following Kerion Celsi – A Follow-up Examination
Foged, E. K. and Jepsen, L. V. (1984), Hair Loss Following Kerion Celsi-A Follow-up Examination. Mycoses, 27: 411–414

Registrars Symposium Case presentations

A newborn referred with blistering

W. Abdelrahman, S Clements, S Hoey
Dept of Dermatology, Royal Victoria Hospital, Belfast, Northern Ireland

Background:
Baby O was born at 38+3 weeks gestation by emergency C-section. From birth he was noted to have non-inflammatory blistering around his legs, hands and flanks; with scaling/ peeling in other areas of his body. His nails were unremarkable and there was no oral mucosal involvement.

Differential Diagnosis:
The presence of blisters and scaling of skin was suggestive of Epidermolytic Ichthyosis. The main differential was epidermolysis bullosa. Of note there was no family history of blistering skin conditions.

Diagnosis:
A skin biopsy was taken and sent to The National Diagnostic Epidermolysis Bullosa laboratory at St John's Institute of Dermatology in London for analysis. Histology appearances were highly suggestive of epidermolytic ichthyosis. The next step was to sequence KRT1 and KRT10 for mutations. Baby O had a missense mutation in keratin 1 which has been previously reported as pathogenic and the cause of this condition. A keratin 10 mutation was also detected.

Bullous ichthyosis:
Bullous ichthyosis, also known as bullous ichthyosiform erythroderma (BIE), is a rare autosomal dominant keratin (either keratin 1 or 10 mutations) disorder occurring in 1 in 300,000 births. It presents with skin fragility leading to blistering, peeling and erosions in the neonate, similar to staphylococcal scalded skin syndrome or epidermolysis bullosa. BIE tends to improve with increasing age. There are no general laboratory studies needed for diagnosis, but keratin defect studies can be performed and once a mutation is identified, mutation specific testing for relatives is available. In addition to presentation and history, skin biopsy is also helpful in aiding diagnosis. Typical histological features include marked hyperkeratosis, thick granular layer, coarse keratohyaline granules and vacuolar degeneration of the upper epidermis. Newborns with BIE are at increased risk of infection, secondary sepsis and electrolyte imbalance, therefore they should be transferred to neonatal ICU for monitoring and treatment. Gentle handling is also important to avoid trauma to the skin. The mainstay of treatment is good wound care for blistering and the use of emollients/antiseptics.

Follow-up
Baby O has done remarkably well since discharge. He has had no new blisters forming and is gaining weight.

Treatment of cutaneous metastatic melanoma with topical diphencyprone - a case report

I.McDonald, F.M. Keane, J.A. McCaffrey, F.J. Moloney
Department of Dermatology, Mater Misericordiae University Hospital

Introduction:
Treatment of metastatic melanoma with contact sensitizers and topical immunotherapy was first reported over 30 years ago. Diphencyprone (DPCP) is a potent contact sensitizer which is sometimes used in the treatment of alopecia areata and cutaneous warts. More recently, case reports have demonstrated its efficacy in the management of cutaneous metastatic melanoma, with its first reported use as a single agent for this purpose in 20071. Its hypersensitivity response is thought to induce a lymphocyte mediated tumour destruction, although the exact mode of action has yet to be determined.

Case History:
A 57-year-old man with a primary superficial spreading melanoma (Breslow thickness 2.3mm, mitotic count 4-5/10hpf), on his left lower back was treated with wide local excision in June 2011. Sentinel node biopsy revealed a single positive inguinal node. He received high dose adjuvant interferon from September 2011 until October 2012. In February 2013 he presented with extensive, pigmented nodules and papules on his right buttock and hip. Histology confirmed locoregional recurrent melanoma. Imaging at that time revealed no distant metastases. He remained clinically well and screening for the BRAF mutation was negative. Multidisciplinary discussion deemed surgical resection or radiotherapy unsuitable and the option of topical immunotherapy was explored.

Treatment regime:
Following consent the patient was sensitized by the application of 2% DPCP in acetone to skin on his upper arm. 5% Imiquimod cream was applied to an area 8x8cm on the affected buttock, on two separate days. Twenty four hours later 0.1% DPCP in aqueous cream was applied and left on for 12 hours. He was reviewed weekly before further application of DPCP. 5% Imiquimod cream was applied on the day prior to review. Because of significant blistering and erythema at the site of application the concentration of DPCP was reduced to 0.01% and Imiquimod was held. After three weeks treatment the cutaneous metastases flattened from baseline. At four weeks, two nodules had resolved and at five weeks further flattening was observed. A decision to discontinue DPCP at that point was taken due to patient discomfort from localised inflammation.

Conclusions:
The management of extensive in transit metastases is difficult and must be individualized and discussed by a multidisciplinary team. Topical DPCP is an inexpensive and generally well tolerated therapeutic option for slow growing cutaneous metastatic melanoma deemed unsuitable for surgery, radiotherapy or targeted therapy.

1 "http://www.ncbi.nlm.nih.gov/pubmed/24522938"Topical immunotherapy with diphencyprone for in transit and cutaneously metastatic melanoma. Damian DL, Saw RP, Thompson JF.J Surg Oncol. 2014

A case of melanonychia in a child.

RH El-khayat1, M Y Walsh2, A Rashid3, K McKenna1 .
[1.] Department of Dermatology [2.] Department of Pathology
[3.] Department of Plastic Surgery. Belfast Health and Social Care Trust.

Case description:
A 2-year old child, presented at the age of 3 months with pigmentation of the left great toe nail. The pigmentation has been present since birth. On examination the following were noted: Brown pigmentation across the nail fold and nail bed, as well as, multiple fine longitudinal pigmented bands.
At clinic review 4 months later, the nail was reported to be brittle and the following changes to the pigmentation pattern were noted: The pigmentation in the proximal nail fold had spread more proximally, a single longitudinal pigmented wide band and pigmentation in the lateral nail fold. Subsequently, a wedge excision of eponychium was arranged. The histopathology report was inconclusive with no melanocytic lesion or melanocytes identified in the specimen.

Digital dermoscopy assessment was then carried out, which showed features in keeping with melanocytic lesion. The case was discussed at the skin cancer MDT and nail bed with nail fold limited transverse excision was recommended. The histopathology report on this occasion confirmed subungual compound naevus with no evidence of dysplasia or malignancy. Following the transverse excision, persistent abnormal pigmentation was noted. Therefore, wider excision of recurrent melanonychia with full thickness graft was performed. The histopathology report confirmed the complete excision of the compound naevus.

Discussion:
The initial wedge biopsy was inconclusive. However, the worrisome changes noted in our case on subsequent clinical reviews warranted both limited and wider excision to exclude malignant transformation arising in a benign precursor.
Subungual congenital nevi are rare with less than 20 biopsy-proven cases reported. All the reported congenital subungual melanocytic nevi had a false positive Hutchinson sign (periungual hyperpigmentation with longitudinal melanonychia).

Reference :
Goldminz AM, Wolpowitz D, Gottlieb AB, Krathen MS. Congenital subungual melanocytic nevus with a pseudo-Hutchinson sign. Dermatol Online J. 2013 Apr 15;19(4):8

Poster Presentations - Running Order

P 01
Epidermal Protease-activated receptor-2 (PAR2) overexpression causes atopic dermatitis-like skin disease: Neuro-Epidermal Communication
T. Buhl1,2, A. Ikoma3, F. Cevikbas3, C. Kempkes3, M. Sulk3, T. Akiyama5, E. Carstens5, M.P. Schön2, P. Elias3, S.R. Coughlin6, and M. Steinhoff1
1 Dept. of Dermatology and UCD Charles Institute for Translational Dermatology, Dublin, Ireland; 2 UMG Dermatology, Göttingen, Germany; 3 UCSF Dermatology, San Francisco, CA, USA; 4 UKM Dermatology, Münster, Germany; 5 UCD Center for Neuroscience, Davis, CA, USA; 6 UCSF Cardiovascular Research Institute, San Francisco, CA, USA.

P 02
Molecular and morphological characterization of the inflammatory infiltrate in Rosacea: new insights into immune pathophysiology
Timo Buhl3,4, Mathias Sulk2, Pawel Nowak2, Jörg Buddenkotte2, Ferda Cevikbas1, Cordula Kempkes1, Jerome Auberts, Johannes J. Voegel5, and Martin Steinhoff4
1Dermatology, University of California, San Francisco (UCSF), San Francisco, CA, USA; 2Dermatology, University of Münster, Münster, Germany; 3Dermatology, University of Göttingen, Göttingen, Germany; 4Charles Institute for Translational Dermatology, University of Dublin, Dublin, Ireland; 5Molecular Dermatology, Galderma R&D, Sophia Antipolis, France

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P 03
Actinic granulomas in a patient with poorly controlled diabetes mellitus: a diagnostic dilemma.
R. Hellen, R. O'Connor, M. Connolly, N. Leonard, AM. Tobin.
The Adelaide and Meath Hospital, Tallaght, Dublin 24

P 04
Behçet's disease occurring within plaques of necrobiosis lipoidica
S. McCarthy, I. McDonald, C. Fahy, P. Lenane
Department of Dermatology, Mater Misericordiae University Hospital

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P 05
A case of aromatase inhibitor - induced lupus erythematosus
S. McCarthy, I. McDonald, C. Fahy, P. Lenane
Department of Dermatology, Mater Misericordiae University Hospital

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P 06
Pregnancy induced rosacea fulminans, what happens in subsequent pregnancies?
R. O'Connor, R. Hellen, M. Connolly, AM. Tobin.
The Adelaide & Meath Hospital, Tallaght, Dublin 24.

P 07
Two cases of significant weight loss on isotretinoin.
R. O'Connor, R. Hellen, M. Connolly, AM. Tobin
The Adelaide & Meath Hospital, Dublin

Poster Presentations - Abstracts

Epidermal Protease-activated receptor-2 (PAR2) overexpression causes atopic dermatitis-like skin disease: Neuro-Epidermal Communication

T. Buhl1,2, A. Ikoma3, F. Cevikbas3, C. Kempkes3, M. Sulk3, T. Akiyama5, E. Carstens5, M.P. Schön2, P. Elias3, S.R. Coughlin6, and M. Steinhoff1
1 Dept. of Dermatology and UCD Charles Institute for Translational Dermatology, Dublin, Ireland; 2 UMG Dermatology, Göttingen, Germany; 3 UCSF Dermatology, San Francisco, CA, USA; 4 UKM Dermatology, Münster, Germany; 5 UCD Center for Neuroscience, Davis, CA, USA; 6 UCSF Cardiovascular Research Institute, San Francisco, CA, USA.

Protease-activated receptor-2 (PAR2) activation has been implicated in the pathophysiology of atopic dermatitis, Netherton syndrome, pruritus, as well as impaired skin barrier regulation. With the aim to study the effects of epidermal PAR-2 function on skin inflammation and itch, we generated a mouse that overexpresses PAR2 in keratinocytes only (KC-PAR2OE). Although KC-PAR2OE newborns display no overt abnormalities, they spontaneously develop dry skin, severe pruritus, and subsequently eczematous skin lesions after several weeks. Analysis of barrier function and immune response in lesional KC-PAR2OE mice revealed the hallmarks of atopic dermatitis-like skin lesions including acanthosis, parakeratosis, significant downregulation of filaggrin and other epidermal structure proteins, a mast cell- and T cell-driven inflammatory infiltrate. Of note, and in close correlation to patients with atopic dermatitis, repeated topical application of house dust mite (HDM) allergens onto KC-PAR2OE mice induced earlier and more severe lesions and pruritus in these mice (as determined by increased skin lesion score, scratching bouts, TEWL, total IgE). Our electrophysiological, morphological and molecular studies show that KC-PAR2OE mice have an increased density of unmyelinated nerve fibers, increased NGF and endothelin expression levels in the skin, which may explain our findings of higher susceptibility of KC-PAR2OE mice to pruritogens and the development of spontaneously increased pruritus. In sum, our results suggest that certain proteases and KC-PAR2 are critically involved in the pathophysiology of pruritus and atopic dermatitis. KC-derived PAR2 seems to be an important link in neuro-epidermal communication with the keratinocyte-protease-PAR2 system as a forefront of sensory signaling and neuro-immune communication in inflammatory skin diseases.

Molecular and morphological characterization of the inflammatory infiltrate in Rosacea: new insights into immune pathophysiology

Timo Buhl3,4, Mathias Sulk2, Pawel Nowak2, Jörg Buddenkotte2, Ferda Cevikbas1, Cordula Kempkes1, Jerome Aubert5, Johannes J. Voegel5, and Martin Steinhoff4
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Rosacea is a common chronic inflammatory skin disease of unknown etiology. Thus, the treatment for rosacea is often only symptomatic and a deeper insight into the exact pathophysiology is needed. Therefore, we performed a detailed transcriptome analysis of relevant genes in rosacea subtypes (ETR, PPR, and PhR), compared to non-lesional or healthy skin. Here, we focused on the genes involved in innate or adaptive immunity, and characterized the inflammatory infiltrate using immunohistochemistry. In all rosacea subtypes, the T cell marker CD3 was increased on gene level. Using immunohistochemistry and morphometry, we found an increased infiltrate of CD4+ T cells in ETR and PPR, in particular T helper 1 (TH1) and T helper 17 (TH17) cells. In contrast, molecular markers for B cell activation were only occasionally increased, and immunohistochemistry confirmed B cells to be only occasionally localized in single patients with PPR or PhR. High expression levels of chemokines and cytokines known to be involved in immune cell recruitment and activation of macrophages and mast cells were also observed. In sum, the immune response in rosacea shows a TH1/TH17 expression profile, although slight differences between each subtype exist. Moreover, B cells are only occasionally observed, indicating different trigger factor leading to rosacea. Of innate immune cells, macrophages and mast cells were abundantly present, neutrophils only in pustules, correlating well with transcriptome data. No differences were found for Langerhans cells, NK cells or basophils in rosacea patients compared to controls. Our data give a better understanding about the underlying immune pathways in the pathophysiology of rosacea that may lead to novel, more specific therapies for this frequent chronic inflammatory skin disease.

Actinic granulomas in a patient with poorly controlled diabetes mellitus: a diagnostic dilemma.

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Actinic granuloma (AG) is an uncommon granulomatous condition which is characterized by annular plaques on actinically damaged, photo-exposed skin.

We report a case of a 48-year-old woman who presented with a five month history of a pruritic photo-distributed, eruption on her face, arms, chest wall and upper back. The patient also had poorly controlled type II diabetes with an elevated haemoglobin A1c (62 mmol/mol; reference range ‹ 53 mmol/mol). She also had a positive antinuclear antibody (ANA) and anti-U1RNP antibodies. Oral and topical steroids had not improved her symptoms.
On physical examination, she had erythematous, infiltrated, annular and arcuate plaques with central clearing in photo-exposed areas. The surrounding skin was photo-damaged.

Histology showed necrobiosis of collagen with surrounding palisading histiocytes, occasional giant cells and a mild perivascular lymphohistiocytic infiltrate. These findings were consistent with a number of differential diagnoses including granuloma annulare, necrobiosis lipoidica, actinic granulomas and interstitial granulomatous dermatitis with arthritis.

The patient was treated as eruptive granuloma annulare and commenced on bath PUVA. After two exposures, she became progressively symptomatic with new lesions and phototherapy was discontinued. This development was more consistent with a diag-

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nosis of actinic granulomas and the patient was commenced on hydroxychloroquine 200mg twice daily with photoprotection. After nine months of treatment, her plaques had flattened with reduced erythema and pruritus.

We report this case as a diagnostic dilemma of actinic granulomas mimicking granuloma annulare in a patient with poorly controlled type II diabetes.

Behçet's disease occurring within plaques of necrobiosis lipoidica

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Abstract:
We describe the first case of behçet's ulceration occurring within plaques of pre-existing necrobiosis lipoidica.

Case History:
A 61 year old lady presented with acute onset of painful lower limb ulceration in the setting of established necrobiosis lipoidica diabeticorum and a 45 year history of beçhets disease.

She was initially diagnosed at the age of 16 when she developed oral and genital ulceration. She has a strong family history of the disease, which affects her sister, brother and daughter. Colchicine treatment was commenced in 2002 to good affect and he developed infrequent flares of the disease up to 2013.

In her early 40's she was diagnosed with necrobiosis lipoidica, in the absence of diabetes mellitus. Erythematous papules developed on her anterior shins bilaterally, without ulceration, and formed atrophic plaques. Topical steroids were commenced initially and subsequently topical tacrolimus was prescribed. In late 2013 she developed breakdown of the areas of necrobiosis lipoidica on her lower limbs. The areas of ulceration were deep and tender with an overhanging inflammatory edge and malodourous discharge. She was prescribed oral erythromycin for ten days, and her colchicine was recommenced. The lesions, however, deteriorated rapidly. Multiple skin biopsies of the inflammatory margin were performed which showed acute inflammatory and vascular changes, and a neutrophilic pustular dermatosis, most consistent with pyoderma gangrenosum-like behçet's ulceration within plaques of necrobiosis lipoidica.

Discussion:
Behçet's disease is a systemic vasculitis of small and large vessels affecting both veins and arteries. It is characterized by recurrent oral aphthae, followed by genital ulcers, skin lesions, arthritis, uveitis, thrombophlebitis, and gastrointestinal and central nervous system involvement. Cutaneous lesions can include pyoderma gangrenosum-like lesions, and pathergy. Necrobiosis lipoidica (NL) is a chronic granulomatous disease. It is usually diagnosed clinically. Erythematous papules develop on the anterior aspect of the lower extremities that can coalesce to form atrophic plaques with telangiectasia. While ulceration is common is NL, the rapid deterioration in this case was unusual. It is possible that pathergy has a role to play in its development within plaques of NL.

A case of aromatase inhibitor - induced lupus erythematosus

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Introduction:
Subacute lupus erythematosus is typically associated with photosensitivity and annular or papulosquamous lesions occurring on sun – exposed skin. Positive anti – Ro antibodies are associated with this disorder, which is also associated with potential drug induction or exacerbation and as a paraneoplastic phenomenon.

Case Report:
We report the case of a 91-year old female, referred to the tertiary referral department of dermatology in 2012. She was being treated for ER positive breast cancer with an aromatase inhibitor, anastrazole and a rash developed two weeks post induction.

The primary lesion was an erythematous annular rash and it affected the face, neck, upper limbs, chest and left breast, which was tender to palpation. Anti - Ro antibody was positive and anti – histone antibody was negative. Initial treatment with oral steroids and antibiotics had a positive response however there was recurrence of the rash on cessation of steroid therapy. The initial differential diagnosis included drug induced lupus erythematosus, erythema multiforme and vasculitis. Histopathology of a biopsy showed features which suggested erythema multiforme or a fixed drug eruption. Clinically however, the rash was most in keeping with drug-induced subacute lupus erythematosus. The aromatase inhibitor was the likely culprit. In early 2013, the patient re-presented with recurrence of subacute lupus erythematosus following initiation of tamoxifen treatment.

Discussion:
The induction of cutaneous lupus by drugs, which are associated with treatment of breast carcinoma, has been previously reported. Tamoxifen was shown to induce subacute lupus in two women attending dermatology services in Nancy, France.1 Trancart et al reported the first case of anastrazole induced subacute lupus in a 73 yr old woman with breast cancer in 2007.2

The development of subacute lupus was thought to primarily be secondary to drug induction due to the chronology of cutaneous disease and commencement of hormonal treatment for breast carcinoma. Considering the rapid development of new, advanced treatments for various carcinomas, further cutaneous adverse effects from these new triggers may be likely. Also it is interesting to note, while oestrogens typically aggravate lupus, tamoxifen and anastrazole are both anti-oestrogens, demonstrating a paradoxical effect.

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Pregnancy induced rosacea fulminans, what happens in subsequent pregnancies?

R. O'Connor, R. Hellen, M. Connolly, AM. Tobin.
The Adelaide & Meath Hospital, Tallaght, Dublin 24.

Rosacea fulminans is a rare facial dermatosis with a female preponderance. It is characterised by the sudden onset of numerous papules, pustules and coalescent nodules localised to the face. Although the aetiology remains unclear, immunological, hormonal and vascular factors have been suggested. To our knowledge there are fifteen reported cases of rosacea fulminans associated with pregnancy, suggesting that hormonal factors may be a trigger. There is no clear evidence as to the mechanism by which pregnancy may trigger this condition. Early and aggressive treatment is essential, yet in the setting of pregnancy a more cautious approach is needed. Effective therapeutic options include retinoids, tetracyclines, anti-androgenic contraceptives and dapsone but all contraindicated in pregnancy. We report two cases of pregnancy-induced rosacea fulminans that were successfully treated post partum with isotretinoin.

Our first patient was a 39-year-old female in her second trimester that presented with a 4-week history of sudden onset severe erythema, pustules and fluctuant nodules affecting her chin and cheeks consistent with a diagnosis of acute rosacea fulminans. She previously suffered from acne vulgaris responsive to tetracyclines. She was treated with oral erythromycin, prednisolone and intralesional triamcinolone throughout her pregnancy, which lead to a significant improvement but still reported intermittent flares. She had an uncomplicated delivery of a healthy boy at term. She was commenced on isotretinoin 20mg daily six weeks post partum. Erythromycin and prednisolone were stopped after a normal ACTH stimulation test. Her skin cleared after 7 months of treatment on isotretinoin (20mg daily for 6 months and 10mg for one month). At six month follow up, her skin remained clear off treatment.

Our second patient is a 32-year-old female who was diagnosed with rosacea fulminans at 17 weeks gestation. She had a history of rosacea, which was well controlled prior to pregnancy. On examination she had inflamed nodules, papules and cysts affecting her face. Treatment with erythromycin, prednisolone and intralesional triamcinolone throughout her pregnancy lead to an improvement but not clearance of rosacea fulminans. She had an uncomplicated delivery of a healthy baby girl at term. Post-partum she was commenced on isotretinoin 10mg daily and continued prednisolone and erythromycin. Her dose was increased to 30mg od and treatment is ongoing with good response. This patient hopes to plan a second pregnancy and there is little in the literature to predict the likelihood of recurrence of her rosacea.

The cause of pregnancy-induced rosacea fulminans is poorly understood and presents a therapeutic challenge for future pregnancies.

Two cases of significant weight loss on isotretinoin.

R. O'Connor, R. Hellen, M. Connolly, AM. Tobin
The Adelaide & Meath Hospital, Dublin

Isotretinoin is the gold standard treatment for nodulocystic acne. The commonly reported side effects include dryness of the skin and mucous membranes, hepatitis, pancreatitis, mood disturbance and myalgia. Isotretinoin previously suspected of increasing the risk of inflammatory bowel disease has since been disproven.1 We report two cases of unexplained significant weight loss on isotretinoin for the treatment of acne vulgaris.

An 18-year man presented with a 5-year history of papulopustular acne affecting his face and back. His past medical history included anaemia and vitamin B12 deficiency. On examination he had scattered papules, pustules and open comedones affecting his back and extensive facial acne with scarring. He had failed 3 tetracycline antibiotics and trimethoprim. He was commenced on isotretinoin 0.25mg/kg daily and at that point weighed 84kg. He tolerated treatment well up to a dose of 0.5mg/kg daily, however suffered mood swings and low mood without suicidal ideation. Treatment was discontinued after 11 months and his weight at that point was 74kg. The notable 10kg weight loss was unintentional and he was systemically well with no gastrointestinal symptoms. On three month follow up, he had gained 4kg and his mood improved off the isotretinoin.

An 18-year-old man presented with a 3-year history of Leeds grade IV nodulocystic acne on the face, back and chest. He had failed antibiotic treatment over two years. His weight was 60kg and he was commenced on isotretinoin 0.25mg/kg daily, which was increased to 0.5mg/kg daily. He completed 7 months of treatment and his acne cleared fully. His weight 1 month prior to stopping isotretinoin was 57kg. This 3kg weight loss was unintentional and he denied any mood disturbance or systemic symptoms during the course of treatment.

These two cases describe previously undocumented weight loss while undergoing treatment with isotretinoin for acne. The mechanism is unclear and while mood disturbance may be a factor in the first case, it was not a factor in the second case. We wish to highlight two cases of weight loss on isotretinoin for likely two differing reasons.

1."http://www.ncbi.nlm.nih.gov/pubmed?term=Etminan%20M%5BAuthor%5D&cauthor=true&cauthor_uid=23426479"Etminan M1, "http://www.ncbi.nlm.nih.gov/pubmed?term=Bird%20ST%5BAuthor%5D&cauthor=true&cauthor_uid=23426479"Bird ST, HYPERLINK "http://www.ncbi.nlm.nih.gov/pubmed?term=Delaney%20JA%5BAuthor%5D&cauthor=true&cauthor_uid=23426479"Delaney JA, HYPERLINK "http://www.ncbi.nlm.nih.gov/pubmed?term=Bressler%20B%5BAuthor%5D&cauthor=true&cauthor_uid=23426479"Bressler B,"http://www.ncbi.nlm.nih.gov/pubmed?term=Brophy%20JM%5BAuthor%5D&cauthor=true&cauthor_uid=23426479"Brophy JM. Isotretinin and risk for Inflammatory bowel disease: a nested case-control study and metanalysis of published and unpublished data. JAMA Dermatol. 2013 Feb;149(2):216-20.

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Treated area should not be covered with occlusive bandages after applying gel. Optimal therapeutic effect can be assessed after approximately 8 weeks. If treated area shows an incomplete response at follow-up, treatment should be carefully re-evaluated and management reconsidered. Clinical data on more than one treatment course of 2 or 3 consecutive days not available. Clinical data on treatment of more than one area not available. Treatment data in immunocompromised patients are not available, but systemic risk not expected since ingenol mebutate is not absorbed systemically. **Contraindications:** Hypersensitivity to any of the constituents. **Precautions and warnings:** Avoid contact with eyes. If accidental exposure occurs, flush eyes with large amounts of water and seek medical care as soon as possible. Eye disorders such as eye pain, eye lid oedema and periorbital oedema expected to occur after accidental exposure. Gel must not be ingested. If accidental ingestion occurs, drink plenty of water and seek medical care. Administration not recommended until skin healed from treatment with any previous medicinal product or surgery. Do not apply to open wounds or damaged skin. Do not use near the eyes, inside nostrils, inside ears or on lips. Local skin responses (LSRs) such as erythema, flaking/scaling and crusting should be expected to occur after application. LSRs are transient and typically occur within one day of treatment initiation and peak in intensity up to one week following completion of treatment. LSRs typically resolve within 2 weeks of treatment initiation when treating face and scalp and within 4 weeks of treatment initiation when treating trunk and extremities. Treatment effect may not be adequately assessed until resolution of LSRs. Ingenol mebutate did not demonstrate any potential for photo irritation or photoallergic effects during studies to assess the effect of UV irradiation. 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References: 1. Lebwohl M et al. *N Engl J Med* 2012;366:1010-1019.

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