AEDV HIGHLIGHTS
24th World Congress of Dermatology (WCD)

10-15 Junio 2019
Milán
Dermatología pediátrica

Dra. Ana Rodríguez Bandera
Atopic dermatitis

Lawrence Eichenfield
Distinct phenotypes with different disease trajectories

6 latent classes were identified

- **Early-onset-early-resolving AD;** associated with male sex
- **Persistent disease** (early-onset-persistent and early-onset-late-resolving) AD; associated with genetic risk factors and parental history of AD
- **Mid-onset-resolving AD;** not associated with FLG, but associated with asthma

→ Demonstrates the potential importance of a stratified medicine approach
Hard water is rich in dissolved salts (calcium/magnesium)

Direct interaction with *FLG* deficiency

Does a water softener decrease eczema prevalence? = SOFTER trial (*ClinicalTrial identifier: NCT03270566*)
First-in-human topical microbiome transplantation with *Roseomonas mucosa* for atopic dermatitis

Ian A. Myles, …, Gulbu Uzel, Sandip K. Datta

*JCI Insight*. 2018;3(9):e120608. https://doi.org/10.1172/jci.insight.120608

- *Roseomonas mucosa* predominant gram –ve bacterium on skin (70% healthy skin colonized, only 20% in patients with eczema)
- 10 adults + 5 children aerosolized culture applied to skin twice weekly for 6-16 weeks
- 65% skin improvement
- No complications or infections

- Coagulase negative staph (CoNS) species with activity against *Staphylococcus aureus* are common on normal skin and rare in patients with AD
- Introduction of CoNS species onto the skin of patients with AD decreased colonization with *Staphylococcus aureus*
- Antimicrobial peptides produced by CoNS were strain specific, highly potent, and selective against *Staphylococcus aureus*

**Microbiome**

**Microbiome**

**Antimicrobials from human skin commensal bacteria protect against *Staphylococcus aureus* and are deficient in atopic dermatitis**

In vitro study of permeation and penetration of Crisaborole 2% when applied with emollients

• Human skin was placed between donor and receptor compartments of a flow-through diffusion cell
• Crisaborole was applied alone, 15 minutes before, immediately after, or 15 minutes after application of an over-the-counter ointment or over-the-counter moisturizing cream.
• Diffusion of crisaborole was measured 2 hourly for 24 hours
• Results: Crisaborole alone or 15 minutes before cream ointment had equal and significantly higher absorption over crisaborole 15 minutes after cream/ointment or crisaborole 15 minutes after cream/ointment

→ Apply active topical preparation alone or before emollient

- Draelos ZD, et al. AAD 2019
Psoriasis

Dr. Amy Paller
• Retrospective study

• 270 children on methotrexate

➢ Folid acid administration 6-7 times per week was more protective protected more against methotrexate-induced gastrointestinal AEs than weekly administration

- 114 children ≥4-<18 years with severe plaque psoriasis
- 16 weeks
- Randomised to ADA (0.8mg/Kg/15 days)/ ADA (0.4mg/Kg/15days)/ MTX (0.1-0.4mg/Kg/7days)
  - ADA (0.8mg/Kg/15 days) showed significant improvements in PASI 75 compared with MTX
  - No new safety risks.


- 108 children ≥4-<18 years with severe plaque psoriasis
- 52 week long-term extension (16 weeks initial treatment/36 weeks withdrawal period/16 weeks re-treatment)
- Randomised to ADA (0.8mg/Kg/15 days)/ ADA (0.4mg/Kg/15days)/ MTX (0.1-0.4mg/Kg/7days)
  - Maintained or improved efficacy.
  - Improvement in MTX non-responders with ADA (0.8mg/Kg/día)
  - No new safety risks.

CAPE
- Early onset
- Prominent involvement of cheeks, chin and ears
- Family history of psoriasis or PRP
- Minimal response to conventional topical and systemic therapies
- Good response to ustekinumab

➢ CARD 14 sequencing
➢ Treatment with ustekinumab
Genodermatoses

Dr. Christine Bodemer
Dr. Michele de Lucas
Prenatal Correction of X-Linked Hypohidrotic Ectodermal Dysplasia

Holm Schneider, M.D., Florian Faschingbauer, M.D., Sonia Schuepbach-Mallepell, Ph.D., Iris Körber, Sigrun Wohlfart, Ph.D., Angela Dick, Ph.D., Mandy Wahlbuhl, Ph.D., Christine Kowalczyk-Quintas, Ph.D., Michele Vigolo, M.S., Neil Kirby, Ph.D., Corinna Tannert, R.A., Oliver Rompel, M.D., Wolfgang Rascher, M.D., Matthias W. Beckmann, M.D., and Pascal Schneider, Ph.D.

- Genetic deficiency of ectodysplasin A (EDA) causes X-linked hypohidrotic ectodermal dysplasia (XLHED) with impaired sweating and a risk of life-threatening hyperthermia.

- A recombinant protein that includes the receptor-binding domain of EDA was administered intraamniotically to twins at gestational age 26 and 31 weeks and to a single fetus at 26 weeks.

- Result: XLHED-related illness did not develop by 22 months follow-up and the subjects were observed to sweat normally.
Topical Diacerein Ointment for Epidermolysis Bullosa Simplex: A Review

By STL Volume 24 Number 3 - May 12, 2019

Allison L. Limmer, BA, BS\(^1\); Crystal E. Nwannunu, BS\(^1\); Radhika Shah, BS, MS\(^2\); Kendall Coleman, BSA\(^1\); Ravi R. Patel, MD\(^3\); Uyen Ngoc Mui, MD\(^3\); Stephen K. Tyring, MD, PhD\(^{1,3}\)

- EBS is due to a mutated Keratin 14 gene and constitutive activation of IL-1

- Topical 1% diacerein, a drug used to stabilize cartilage in osteoarthritis, has been shown to be an effective and safe in treatment in this condition

- It has been granted USA FDA Rare Pediatric Disease designation in may 2018

- ➔ A new use for an old drug
Long-Term Stability and Safety of Transgenic Cultured Epidermal Stem Cells in Gene Therapy of Junctional Epidermolysis Bullosa

Laura De Rosa,1,5 Sonia Carulli,1,5 Fabienne Cocchiarella,1 Daniela Quaglino,2 Elena Enzo,1 Eleonora Franchini,1 Alberto Giannetti,3 Giorgio De Santis,4 Alessandra Recchia,1 Graziella Pellegrini,1 and Michele De Luca1,6,*

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Correction of Recessive Dystrophic Epidermolysis Bullosa by Transposon-Mediated Integration of COL7A1 in Transplantable Patient-Derived Primary Keratinocytes

Maria Carmela Latella1, Fabienne Cocchiarella1, Laura De Rosa1, Giandomenico Turchiano1,4, Manuel A.F.V. Gonçalves2, Fernando Larcher3,4,5, Michele De Luca1 and Alessandra Recchia1
Infectious dermatosis

Dra. Iria Neri
Congenital Herpes Simplex Virus Type 1 Infection Mimicking Congenital Erosive and Vesicular Dermatosis: Possible Pathogenic Insights

Andrew W. Swarbrick, M.B.B.S.,* Christopher C. Blyth, M.B.B.S. (Hons.),†,‡ and Anne R. Halbert, M.B.B.S. (Hons.)*

- HSV1 infection isolated with PCR in a patient

- Authors note 20% recurrent HSV infection in first year of life in previously reported cases

??Are at least some previous cases of CEVD undiagnosed congenital HSV infection??
Multiple neonatal staphylococcal cold abscesses in large skin folds: a benign neonatal skin infection

• Retrospective observational study from 28 dermatology departments (13 cases)

• Localization in large skin folds, absence of inflammation or fever

• Regardless of treatment, all had a favorable outcome