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CONFERENCE UPDATE

 2023 European Society for Pediatric Dermatology

ABCA12 congenital ichthyosis, from acral abnormalities to systemic manifestations: A syndromic ichthyosis?



CROSSWORD & QUIZ

Think Moisturizer
Think Ajanta Dermatology

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Facial moisturiser



60 / 100 g Cream Aquaxyl Uvinul A Plus, Uvinul T 150, Tinosorb S, Vitamin E

Aqurea HF 20 10 Urea 40% Urea 20% Urea 10%

Ceramide & Colloidal Oat meal based moisturiser

Biosik

Carame 1.344, Ou'Con, Provesto

Cation (Caragon)



Ultra moisturising complex

SORILAST

Ures 12%, Salicylic acid
0.5%, Lactic acid 6.81%,
Avena Sativa 0.10%, HACE 200 0.10%,
Witch Hazel extract 1%, Biophilic HMB 1%



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ISSUE 5





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Ceramides and xerosis

Ceramides are the main intercellular lipids in the horny layer of skin, accounting for 40 to 50 percent of total lipids. There are nine ceramide subclasses in the stratum corneum, each one a combination of a fatty acid and a sphingoid base. Ceramides play an important role in maintaining skin barrier function. These changes in

ceramide levels and profiles result in altered lipid packing exhibited by less ordered lipid structures, which has been identified as a cause of increased stratum corneum permeability. Changes in ceramide composition have also been reported in seasonal and age-related xerosis.¹

Ceramides in skin barrier function and hydration

- Ceramides are the major lipid constituent of lamellar sheets present in the intercellular spaces of the stratum corneum. These lamellar sheets are thought to provide the barrier property of the epidermis.
 Ceramides play an essential role in structuring and maintaining the water permeability barrier function of the skin.² Stratum corneum chymotryptic enzyme
- levels are reduced in the outer layers of the xerotic stratum corneum compared with normal skin.³
- Ceramides help to seal the gaps between skin cells, preventing water from evaporating from the skin and have a strong ability to bind water molecules and maintain skin hydration by forming a network structure in the stratum corneum.²⁴⁵

Ceramides for xerosis

- Basic skin care in the treatment of xerosis cutis is intended to improve skin hydration, compensate for the lack of barrier lipids, and improve the skin's barrier function. Thus, a combination of hydrophilic and lipophilic components is preferable.⁶
- Applications of the stratum corneum lipids cause a significant recovery in either conductance value or scaling, the ceramide fraction being the one that induces the highest increase in conductance.²
- Recent instrumental investigations on xerotic skin support the significance of ceramides in maintaining skin health. Through techniques such as confocal Raman microspectroscopy and high-performance
- liquid chromatography, researchers have demonstrated, *in vivo* and non-invasively, the disorganization and reduction in the concentration of ceramides in xerotic skin samples.⁷
- Based on scientific evidence, ceramides have been incorporated into dermato-aesthetic products to improve skin barrier studies and replenish skin ceramides externally.⁷
- Several studies have shown that emollients containing ceramides can provide superior hydration to the skin compared to other preparations, offering additional benefits to the structure and function of the skin barrier.⁷

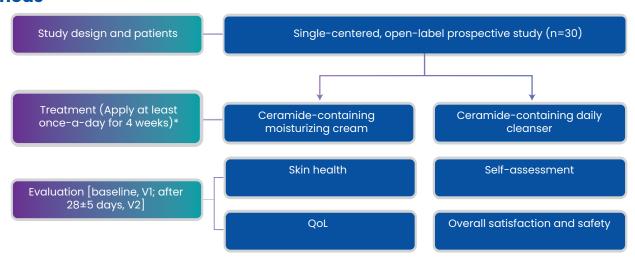




Ceramide-containing skincare routine for elderly patients with xerosis⁷

Researchers conducted a prospective open-label trial to evaluate the effectiveness of a ceramide-based moisturizing cream and cleanser routine on elderly (age ≥70 years) xerosis.

Methods



QoL: Quality-of-life; V1: Visit 1; V2: Visit 2.

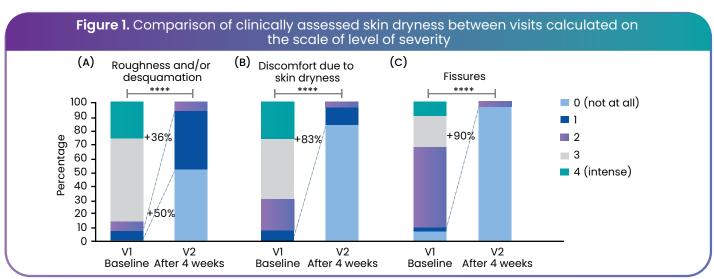
*Specifically, the two products contained bioidentical synthetical ceramides 1, 3 and 6-II as main active ingredients, formulated in a time-released Multilamellar Vesicular Emulsion (MVE) technology.

Results

Skin health

After the 4-week treatment, the symptoms of diseases were significantly ameliorated:

- At visit 2 (V2), the degree of roughness or desquamation showed a median difference in obstructed defecation syndrome (ODS) score of -3.0 (p<0.0001), with most subjects classified as 0 or 1 (Figure 1A).
- Similarly, the second evaluation revealed a significant decrease in discomfort due to skin dryness and in the severity level of the fissures, with a median difference in ODS scores of -3.0 (p<0.0001) and -2.0 (p<0.0001), respectively.
- At the end of the treatment, 83% of patients reported no discomfort, and 97% of patients had no fissures (Figures 1B and 1C, respectively).



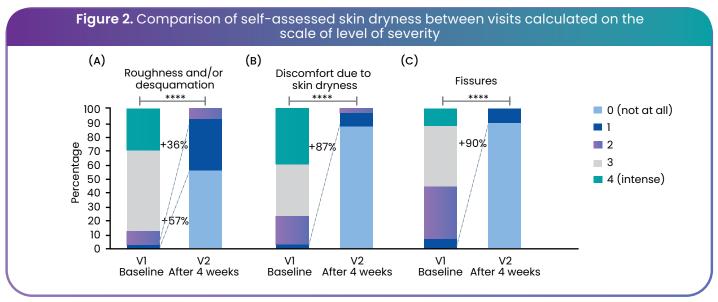




Self-assessment

The results of the self-assessment on skin dryness severity before and after the treatment showed:

- Significant self-perceived improvement in skin roughness/desquamation, with an 85% decrease in symptom severity measured between Visit 1 (V1) and V2 (p<0.0001; Figure 2A).
- A significant decrease in discomfort due to skin dryness was reported, with an overall reduction of 96% in the degree of discomfort at V2 (p<0.0001; Figure 2B).
- Fissures or crackling showed the best improvement after treatment, with a mean reduction of 97% (p<0.0001; Figure 2C) in the severity score.



^{****}p<0.0001. V1: Visit 1; V2: Visit 2.

Quality-of-life (QoL)

The QoL has globally improved in all patients, with a significant reduction of embarrassment and a positive impact on social and daily activities.

Overall satisfaction and safety

Patients' overall satisfaction was high in 87% of patients, and 93% of them stated that they would continue the treatment after the study had been completed which suggests that the treatment was well-tolerated by the patients, further dermatological QoL would be better profile.

After 4 weeks, the daily use of a ceramide-based routine significantly improved signs and symptoms of senile xerosis and chronic discomfort associated with xerosis in elderly adults.

References: 1. Weber TM, Kausch M, Rippke F, et al. Treatment of xerosis with a topical formulation containing glyceryl glucoside, natural moisturizing factors, and ceramide. J Clin Aesthet Dermatol. 2012;5(8):29–39. 2. Coderch L, López O, Maza A, et al. Ceramides and skin function. Am J Clin Dermatol. 2003;4(2):107–129. 3. Rawlings AV, Matts PJ. Stratum corneum moisturization at the molecular level: An update in relation to the dry skin cycle. J Invest Dermatol. 2005;124(6):1099–1110. 4. Nafisi S, Maibach H I. Nanotechnology in cosmetics. Cosmetic Science and Technology. 2017;337–369. 5. Ceramide metabolism: Structure, functions, and analysis. Available at: https://www.creative-proteomics.com/resource/ceramide-metabolism-structure-functions-and-analysis.htm#:-:text=Ceramides%20have%20a%20strong%20ability,moisturizing%20effect%20on%20the%20skin. Accessed on November 22, 2023. 6. Augustin M, Wilsmann-Theis D, Körber A, et al. Diagnosis and treatment of xerosis cutis - a position paper. J Dtsch Dermatol Ges. 2019:17 Suppl 7:3–33. 7. Filippi F, Chessa MA, Bardazzi F, et al. An easy-to-use, ceramide-containing skincare routine: Effectiveness and improvement of quality of life in elderly patients with xerosis. Ital J Dermatol Venerol. 2023;158(6):429–436.







GUIDELINE/CONSENSUS CORNER

Lamellar ichthyosis guidelines: Consensus recommendations for the use of retinoids in ichthyosis and other disorders of cornification

- Topical and systemic retinoids have long been used in the treatment of ichthyoses and other disorders of cornification.
- Due to the need for long-term use of retinoids for these disorders, often beginning in childhood, numerous clinical concerns must be considered.
- Systemic retinoids have known side effects involving bone and eye. Additionally, potential psychiatric and
- cardiovascular effects need to be considered.
- Contraceptive concerns, as well as the additive cardiovascular and bone effects of systemic retinoid use with hormonal contraception, must also be deliberated for patients of childbearing potential.
- The consensus statements on the effects of retinoids on the skin are explained in Table 1.

Table 1. The consensus statements on the effects of retinoids on the skin					
Consensus statements	SORT				
Both topical and systemic retinoids can improve scaling in patients with select forms of ichthyosis.	IIB				
In generalized disorders that feature prominent scales, there is evidence for the utilization of retinoids. Subtypes that have shown improvement with the use of oral or topical retinoids include:					
Congenital ichthyosiform erythroderma (select genotypes), epidermolytic ichthyosis, erythrokeratodermia variabilis, harlequin ichthyosis, ichthyosis with confetti, IFAP syndrome, KID syndrome, KLICK syndrome, lamellar ichthyosis, loricrin keratoderma, neutral lipid storage disease with ichthyosis, recessive X-linked ichthyosis, and Sjögren-Larsson syndrome.	IIB				
Subtypes for which there is either no data for the use of retinoids or there has been data showing no improvement include:					
CHILD syndrome, CHIME syndrome, Conradi-Hünermann-Happle syndrome, ichthyosis-hypotrichosis syndrome, ichthyosis hypotrichosis-sclerosing cholangitis, ichthyosis prematurity syndrome, MEDNIK syndrome, peeling skin disease, Refsum syndrome, and trichothiodystrophy.	IIIC				
Utilization of retinoids in some disorders with skin fragility, peeling skin, atopic diathesis, or excessive desquamation (e.g., Netherton syndrome) may exacerbate disease and should be used with caution.	IIB				
Both adults and children with moderate-to-severe disorders of keratinization with significant functional or psychological impairment should be offered the opportunity to make a benefit/risk assessment of treatment with a systemic.	IIIC				

B: Recommendation based on inconsistent or limited quality patient-oriented evidence; C: Recommendation based on consensus, opinion, case studies, or disease-oriented evidence; CHILD syndrome: Congenital hemidysplasia with ichthyosiform erythroderma and limb defects syndrome; IFAP syndrome: Ichthyosis follicularis, alopecia, and photophobia syndrome; KID syndrome: Keratitis ichthyosis deafness syndrome; KILCK syndrome: Keratosis linearis with ichthyosis congenita and sclerosing keratoderma syndrome; MEDNIK: Mental retardation, enteropathy, deafness, neuropathy, ichthyosis, keratodermia; SORT: Strength of recommendation taxonomy; II: Limited quality patient-oriented evidence; III: Other evidence including consensus guidelines, opinion, case studies, or disease-oriented evidence.

Reference: Zaenglein AL, Levy ML, Stefanko NS, et al. Consensus recommendations for the use of retinoids in ichthyosis and other disorders of cornification in children and adolescents. Pediatr Dermatol. 2021;38(1):164–180.





CONFERENCE UPDATE

2023 European Society for Pediatric Dermatology

ABCA12 congenital ichthyosis, from acral abnormalities to systemic manifestations: A syndromic ichthyosis?

Moghadam P, Bellon N, Boccara O, et al.

Objective



The phenotype of ABCA12 autosomal recessive congenital ichthyoses (ARCI) ranged from mild xerosis to a lethal form of Harlequin ichthyosis. While ABCA12 protein is expressed in extracutaneous tissues such as tonsils, breasts, esophagus, and pituitary glands, ABCA12 ichthyosis is classified in the non-syndromic subtypes. The aim of the study was to analyze whether ABCA12 ichthyosis should be considered as syndromic.

Methods

Retrospective study (n=17)

Data from three reference centers between January 2000 and May 2022

Results

- At birth, the patients presented with superficial desquamation (1/17, 6%), collodion baby (12/17, 71%) and harlequin ichthyosis (n = 4/17, 24%) among whom one patient died.
- At last medical consultation, skin involvement was reported as severe in 4/16 patients (25%), moderate in 7/16 patients (44%) or mild in 5/16 patients (31%). 12/16 (75%) patients had associated erythema.
- Palmoplantar keratoderma was reported in all patients, while 11 (69%) presented with abnormalities of
- the extremities (9 digital retractions, 10 interdigital synechiae with tapered fingers, 5 ulnar deviations, 1 absence of a third phalange).
- Digital retractions were not associated with skin inflammation (p=1), or with severe skin involvement (p=0.162).
- Growth retardation was reported in 9 at birth and in 6 at the last medical visit.
- Insulin-like growth factor 1 (IGF1) was low (2/2), one patient had type one diabetes mellitus, and 10 patients had psychomotor retardation.

ABCA12 ichthyosis might be considered syndromic ichthyosis with intra- and interfamilial variability. It associates skin, joints, and musculoskeletal abnormalities, endocrine glands involvement such as pancreas or hypophysis, and growth retardation.

Reference: Moghadam P, Bellon N, Boccara O, et al. ABCA 12 congenital ichthyosis, from acral abnormalities to systemic manifestations: A syndromic ichthyosis? Pediatr Dermatol. 2023;40(Suppl. 2):10-89.





CROSSWORD & QUIZ

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- The primary function of ceramides in the skin is
 maintenance
- 6. The skin layer which is primarily affected by xerosis is
- 7. The ceramidesgaps between skin cells in the maintenance of the skin barrier

DOWN

- The layer of the skin, which is the outermost and directly impacted by xerosis, is
- 3. In xerosis, ceramides' significance lies in maintaining skinthrough binding water molecules
- 4. In xerosis, ceramide levels in the stratum corneum
- 5. Consequences of compromised skin barrier in xerosis are.....

Down: J. Epidermis; 3. Hydration; 4. Decreases; 5. Infections

Answers: Across: 2. Barrier; 6. Stratum corneum; 7. Seal

- 1. How do ceramides contribute to skin hydration in xerosis?
- A. By promoting oil production
- B. By forming a network structure in the stratum corneum
- C. By enhancing collagen synthesis
- D. By reducing melanin production
- 2. What is the role of ceramides in the lamellar structure of the stratum corneum?
- A. Enhancing pigmentation
- B. Sealing gaps between skin cells
- C. Stimulating hair growth
- D. Reducing sebum production

- 3. Which of the following is a consequence of compromised skin barrier in xerosis?
- A. Increased water retention
- B. Reduced water loss
- C. Enhanced UV protection
- D. Increased susceptibility to infections
- 4. How do ceramides help prevent water loss from the skin?
- A. By forming a protective film on the skin
- B. By reducing sweat production
- C. By constricting blood vessels
- D. By promoting skin peeling