

# Lamellar Ichthyosis: A Case Study

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## BACKGROUND

Lamellar ichthyosis is an autosomal recessive disorder that is apparent at birth and is present throughout life. Patients with lamellar ichthyosis have accelerated epidermal turnover with proliferative hyperkeratosis, in contrast to retention hyperkeratosis. The prevalence of lamellar ichthyosis is less than 1 per 300,000 live births and is equally distributed between males and females. The newborn is born encased in a collodion membrane that sheds within 14 days, revealing generalized scaling with variable redness of the skin. The scaling may be fine or platelike, resembling fish skin. In the neonatal period, following the shedding of the collodion membrane, the newborn is at risk for secondary sepsis and hypernatremic dehydration. As the child ages, the hyperkeratosis can interfere with normal sweat gland function, which can predispose to heat intolerance and possible heat shock. Although the disorder is not life threatening, it can be disfiguring and cause considerable psychological stress.

Since this disorder has no cure, treatment is directed at decreasing symptoms. Emollients are traditionally applied after showering or bathing. The stratum corneum can absorb 6 times its weight in water, and a heavy emollient, such as petrolatum jelly or water-in-oil preparations should be applied while the skin is still wet. Alpha-hydroxy acids, such as lactic acid, help reduce corneocyte adhesion and decrease the thickness of the epidermis. Urea creams can help soften scales. Salicylic acids in combination with propylene glycol help to remove dark scaling. Topical retinoic acids decrease thickened scaling. Because of the significant long-term adverse effects, reserve systemic retinoids for severe disease that is refractory to conventional therapy. Antiseptics and antimicrobials can be used topically to control odor. Many of the medications used for lamellar ichthyosis have a long list of potential adverse effects, and care must be taken to discuss the advantages and disadvantages of treatment. Salicylate toxicity is reported, due systemic absorption of topical salicylic acids, in the treatment of children with ichthyosis. Topical tacrolimus, a macrolide immunosuppressant, should be used with caution because significantly elevated systemic tacrolimus levels have been reported in a patient with lamellar ichthyosis.

Historically, with Lamellas Ichthyosis many topical products have been utilized, all have some positive results, none has proven to be a panacea. Some of the newer therapies that have resulted in clinical improvement are Locobase fatty cream, (5% lactic acid and 20% propylene glycol in lipophilic cream); topical N-acetylcysteine, (an antiproliferative effect), tazarotene topical 0.05%, (a receptor-selective retinoid), and calcipotriol (a synthetic derivative of vitamin D-3). Also new to the market are the Olivamine\* based products. They are a new advanced skin care line that provides the skin with surfactant-free cleansing ingredients, amino acids, vitamins, and antioxidants in addition to the most sophisticated silicones (moisturizers) available for superior deep repair and moisturization.

## PATIENT RECOMMENDATIONS

- Shower only twice a week. Bathe the arm pits and groin more frequently, if necessary.
- Use only tepid (lukewarm) water when showering and limit the time actually in the water.
- Do not use soap, only a phospholipid based cleanser.
- Use a humidifier for your home.
- Consider moving to a warm and more humid climate.
- Consider taking supplements such as Flaxseed Oil, Omega 6 and Omega 3 essential fatty acids and Evening Primrose Oil. This can help provide internal healthy oil for your skin.
- Consider taking an over-the-counter antihistamine for intense itching.

## STUDY PROTOCOL

1. Cleanse with Olivamine based 4-in-1 cleansing lotion or 4-in-1 body cleanser and gently pat dry.
2. Follow immediately with a thin coating of Olivamine based nourishing repair cream and allow to dry.
3. Then apply Olivamine based dimethicone/silicone barrier cream to seal in the moisture and prevent dehydration of the skin.

## CASE STUDY

LP is a 17 year old, Hispanic female who was diagnosed with Lamellar Ichthyosis at birth. She is a high school student and active in young adult church functions. Her lifestyle has been limited in the summer due to extreme heat which was exaggerated by the preparations she was using prior to her participation in the study. She suffered heat exhaustion once due to her condition. She has no other significant medical history. Previous treatment regimen included daily showers or baths followed with application of one of several emollients, such as the alph-hydroxy acids or the water in oil preparations. This provided limited comfort and relief for her.

On presentation, her skin appeared dry and was dry to the touch. She had generalized erythema, flaky and scaling skin. The scaling was most severe on her lateral thighs and lower legs while her shoulders presented with dry flaky skin.

After the first week of the Olivamine based treatment protocol, there was noted improvement in her skin. The erythema and dryness had decreased as well as the scaling. She related that "it felt like my skin could breathe, it didn't feel as hot as before". There was continued improvement at week two. At week 5 after twice daily application her skin showed vast improvement with nearly normal skin appearance, texture and hydration.



Left Shoulder, 8-4-05



Left Shoulder, 9-15-05



Right Shoulder, 8-4-05



Right Shoulder, 9-15-05



Left Thigh, 8-4-05



Left Thigh, 9-15-05



Right Thigh, 8-4-05



Right Thigh, 9-15-05

## RESULTS

The Olivamine based skin care regimen proved to be an excellent option for LP. This pleasant 17 year old describes gaining her life back again. The appearance of her skin and the overall quality of life has improved dramatically.

She relates, "I started using (Olivamine-based) products in mid-August. I mainly used an (water-in-oil base product) since I was about a year old and was very used to it and the way it made my skin feel. While it didn't cure my Ichthyosis, it did provide major relief. It was not without problems though, mainly it was so thick and because I don't have sweat glands, it caused me to get hotter quicker. I could barely go outside in the summer without feeling overheated. Since I started using the (Olivamine-based) products not only have I seen great improvements in the way my skin looks and feels, the products soaks in faster and is a lot more convenient than (my old regimen). I continue to see these improvements everyday and know that eventually my skin will look and feel almost normal as I continue with this treatment."

## REFERENCES

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