

SAFETY AND EFFECTIVENESS OF TOPICAL 10 % N-ACETYLCYSTEINE IN 5% UREA O/W EMULSION FOR CONGENITAL LAMELLAR ICTHYOSIS AND EPIDERMOLYTIC ICTHYOSIS IN CHILDREN

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BACKGROUND

Lamellar ichthyosis is a chronic, disabling disorder with significant effect on quality of life. Treatment of lamellar ichthyosis is based on disease severity, although a permanent cure may not yet be possible. Treatment options for lamellar ichthyosis include topical formulations (classically emollient creams, ointments, keratolytic agents and bath oils) and oral retinoids.

OBJECTIVES

To determine safety and effectiveness of topical 10 % N- acetylcysteine (NAC) in 5% urea emulsion by two cases of congenital lamellar ichthyosis and four cases of epidermolytic ichthyosis.

METHODS

Case 1 and 2: 9 and 12 years old patients with congenital lamellar ichthyosis, extended cutaneous xerosis with dark and medium-sized flakes at upper and lower limbs and sides. One of them with moderate affectation at skin folds. Cases 3, 4, 5 y 6: 16, 17 y 18 months and 10 years old patients with epidermolytic ichthyosis (1, 7 and 9 exon mutation of KRT10 gene). Case 3 presents denudated areas at gluteus, trunk and lower limbs, with subsequent healing and keratotic appearance. Case 4 presents big denudated areas at thorax and white keratotic appearance at palm and at sole, progressing to totally body erosion and completely denudation. Case 5 and 6 presents hyperkeratotic lesion at upper and lower limbs. All patients have been previously treated with emollient creams and ointments.

RESULTS

It was decided to apply topical 10 % N- acetylcysteine in 5% urea emulsion at one limb two times a day for 6 weeks and compare its efficacy to that of an emollient prescription of vaseline, paraffin and glycerol. For sensitive areas (palm, sole and face) the concentration was modified to 5% NAC in 5% urea emulsion presenting better tolerance. The first four cases presented clinical improvement and reduction of the hyperkeratotic lesion without side effects, therefore was treated all the body surface area. Case 5 interrupted the treatment after a month due to a lack of answer and started oral acitretin treatment. Case 6 stopped the treatment because of the emulsion's unpleasant smell.

Case 3 evolution:



CONCLUSION

Even though a larger group of patients should be necessary to confirm data, topical 10 % N- acetylcysteine in 5% urea emulsion seems to be an effective and safety option to reduce hyperkeratotic lesion when emollient creams and ointments aren't effective and before use systemic treatments which could increase the risk of side effects.