

# OFF-LABEL USE OF USTEKINUMAB IN NONBULLOUS CONGENITAL ICTHYOSIFORM ERYTHRODERMA: A CASE REPORT

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## BACKGROUND AND IMPORTANCE

**Nonbullous congenital ichthyosiform erythroderma (NBCIE)** is a rare disease that predominantly affects the skin, courses with palmoplantar keratoderma anhidrosis, and nail dystrophy. In severe cases, there is alopecia in certain areas

## AIM AND OBJECTIVES

The objective of this report is to describe the **off-label use of ustekinumab** in a patient with NBCIE, and to assess the safety of the treatment and tolerability profile

## MATERIAL AND METHODS

- 6 year old man with NBCIE
- Very itchy generalized NBCIE affecting his quality of life

Several **treatment with no success**:  
emollients emulsions, topical and oral corticosteroids and H1-antihistamines

- ✓ Dermatology department to request the off-label use of Ustekinumab
- ✓ The Pharmacy and Therapeutics Committee approved the treatment initiation based on the published efficacy studies. **NBCIE patients have a dominant profile of TH17/IL-23** and it has previously been demonstrated the successful use of ustekinumab in patients affected by ichthyosis
- ✓ Efficacy was evaluated based on **IASI scheme (Ichthyosis Area Severity Index)**

## RESULTS

May 2021 dosis of 1,2 mg/kg of **Ustekinumab**

- The first 2 months, patient received monthly doses while hereafter the dosing was adjusted to a bi-monthly frequency.
- At the **3rd month** of treatment, the patient observed a **mild improvement** while presenting diffused erythema with scaling and rough skin in the folds, with an impact of 60% of body surface.
- At **month 6**, a fourth dose of 27,6mg of ustekinumab weight-adjusted was provided. Thereafter, the patient observed a significant improvement that directly affected his quality of life, **achieving a 60% reduction extension according to IASI and a 66% reduction erythema according to IASI** and no side effects were reported.

Nevertheless Dermatology department request the off-label use of **Dupilumab**

## CONCLUSION AND RELEVANCE

Results demonstrate ustekinumab provides only **partial response** at month 6 of NBCIE treatment, with modest but significant improvement in patient's quality of life with a good safety profile.

