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.

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.

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

✓ 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: dose 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.