

**OFF-LABEL USE OF USTEKINUMAB IN REFRACTORY EPIDERMOLYTIC ICHTHYOSIS: A CASE REPORT**

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**BACKGROUND AND IMPORTANCE**

Epidermolytic ichthyosis (EI) is a skin genetic disorder that predominantly affects joints and friction areas. It is characterised by presenting erythroderma, bad smell, large erosions, hyperkeratosis, recurrent skin infections and itching.

For patients with a mild-moderate disease, topical (keratolytic and emollients) medication is considered the first-line treatment. However, oral retinoids could be a safe and effective choice for some patients who have severe and extensive scaling and hyperkeratosis. In some severe EI subtypes, the off-label use of ustekinumab is based on marked elevation of cytokines in the Th17/IL-23 pathway, similar to inflammatory diseases like psoriasis.

**AIM AND OBJECTIVES**

To evaluate the efficacy and security of ustekinumab in a 8-year-old female patient with severe EI with KRT1 mutation, refractory to topical treatment and oral retinoids.

**MATERIALS AND METHODS**

Firstly, she underwent keratolytics and emollients with scarce clinical response. Due to the absence of effectiveness of these therapies, she received oral retinoids (acitretin) up to maximum tolerated dose, but some erythematous lesions and hyperkeratosis rapidly appeared on her skin. For this reason, the treatment had to be discontinued several times. In April 2022, Dermatology Service requested the off-label use of ustekinumab with a dosage of 0.75 mg/kg at weeks 0,4,8 and 12, and then administered each 12 weeks to the Pharmacy therapeutics committee.

Afterwards, an extensive review was carried out by Pharmacy Service. A report was made with a positive assessment for approval of treatment. This decision was supported by some case reports showing clinical results of ustekinumab in some EI subtypes and the lack of available alternative therapies in this case.

**RESULTS**

At the beginning of treatment with ustekinumab, extensive scaly erythematous lesions with circinate margins were observed, affecting the facial area, trunk and extremities, accompanied by diffuse palmoplantar keratoderma.

After three months of the first administration of ustekinumab, the patient was examined by a dermatologist. An excellent clinical response was observed with resolution of the facial lesions and almost complete on the trunk, with hyperkeratotic lesions persisting in folds, without underlying erythema. Moreover, no adverse events related to ustekinumab were registered.

**CONCLUSIONS AND RELEVANCE**

Ustekinumab has been suggested to be an alternative therapy in some severe EI subtypes refractory to topical and systemic treatments. In spite of being safe and effective in this patient, longer studies are needed to consider ustekinumab in the therapeutic management of EI.

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